RHEUMATOLOGY

MULTIPLE CHOICE QUESTIONS WITH EXPLANATIONS

Dr P A Shah

EDMEDSUR

ISBN: 9781483538112

RHEUMATOLOGY SECTION ONE - APPROACH TO RHEUMATOLOGICAL DISORDERS

Question 1.

A 33-year-old woman presents with a malar rash that is exacerbated by sun exposure. She has experienced episodes of myalgia, pleural effusion, pericarditis, and arthralgia without joint deformity over the course of several years. She has a history of hematuria and no history of drug intake prior to the onset of these symptoms. The best screening test for her disease would be: (AI)

1. Antinuclear antibody
2. Anti-ds-DNA antibody
3. Anti-RNP antibody
4. Anti-histone antibody

DISCUSSION:

A malar rash with arthralgia, serositis, and hematuria in a female of childbearing age suggests the possibility of systemic lupus erythematosus (SLE). However, an SLE-like picture may be seen in drug-induced lupus and mixed connective tissue disorder. Antinuclear antibody is the most sensitive test to screen for SLE. Its repeated absence virtually rules out the possibility of SLE. Anti-ds-DNA (and anti-Sm) antibodies have high specificity and are used to confirm the diagnosis of SLE; however, because of their poor sensitivity, they are not used as screening tests. Mixed connective tissue disorder (MCTD) has features of disorders such as rheumatoid arthritis, limited cutaneous scleroderma (skin stiffness, dysphagia), and polymyositis, in addition to those of SLE. Anti-RNP antibodies are detectable in less than half of SLE patients, but high titers of anti-RNP antibodies are diagnostic of MCTD. Anti-histone antibody is seen in drug-induced lupus (which is unlikely, as there is no history drug intake prior to the onset of symptoms).

ANSWER:

The correct answer is “antinuclear antibody.”

REFERENCES:

Rahman A, Isenberg DA. Chapter 19.11.2. Systemic lupus erythematosus and related disorders. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Hahn B. Chapter 319. Systemic lupus erythematosus. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 2.

A 40-year-old female, with a known case of asthma for the last four years, presented with a two-month history of numbness in the right upper and both lower limbs. Examination revealed asymmetric neuropathy and palpable purpura over the lower limbs. Investigations revealed eosinophilia. What is the likely diagnosis? (PGI)

1. Systemic lupus erythematosus (SLE)
2. Polyarteritis nodosa (PAN)
3. Giant cell arteritis (GCA)
4. Churg-Strauss syndrome

DISCUSSION:

Peripheral neuropathy is broadly divided into mononeuritis simplex, mononeuritis multiplex, and polyneuropathy. This patient has asymmetric neuropathy or mononeuritis multiplex. Common causes of mononeuritis multiplex are vasculitic syndromes, infectious diseases such as HIV or leprosy, and other conditions such as diabetes. A vasculitic neuropathy (palpable purpura) with asthma and eosinophilia favors the diagnosis of Churg-Strauss syndrome. Asthma and eosinophilia are not seen in conditions such as SLE, GCA, or PAN.

ANSWER:

The correct answer is “Churg-Strauss syndrome.”

REFERENCES:

Langford CA, Fauci AS. Chapter 326. The Vasculitis Syndromes. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.
Wells AU, M. du Bois R. Chapter 18.11.5. The lung in vasculitis. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 3.

A five-year-old girl presents with fever and conjunctivitis. Physical examination is significant for oral erythema and fissuring along with a generalized maculopapular rash and cervical lymphadenopathy. What is the most likely diagnosis? (UPSC)

1. Henoch-Schonlein purpura
2. Polyarteritis nodosa
3. Kawasaki disease
4. Takayasu's arteritis

DISCUSSION:

Symptoms of vasculitis vary greatly and depend upon the organs affected and the severity of the disease. The involvement of large vessels (giant cell arteritis, Takayasu's disease) frequently results in limb claudication, asymmetric blood pressure in the limbs, and the absence of pulses. The involvement of medium vessels (polyarteritis nodosa, Kawasaki disease) results in cutaneous nodules, livedo reticularis, microaneurysms, and mononeuritis multiplex. The involvement of small vessels (ANCA-associated vasculitis, Henoch-Schonlein purpura) results in glomerulonephritis, purpura, and alveolar hemorrhage. Kawasaki disease often begins with fever that is not very responsive to paracetamol. Bilateral conjunctival injection usually begins shortly after the onset of fever. It is not purulent, and it is not painful. Oral manifestations of this disease include erythematous and swollen lips and "strawberry tongue." Cervical lymphadenopathy is seen in about three-fourths of patients, which are usually non-tender and non-suppurative. Henoch-Schonlein purpura presents with abdominal pain, rashes, palpable purpura, and arthritis. Polyarteritis nodosa and Takayasu's arteritis have distinct presentations and are unlikely at this age.

ANSWER:

The correct answer is "Kawasaki disease."

REFERENCES:

Langford CA, Fauci AS. Chapter 326. The Vasculitis Syndromes. In: Longo DL, Fauci AS, Kasper

DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Kawasaki T (1967). "Acute febrile mucocutaneous syndrome with lymphoid involvement with specific desquamation of the fingers and toes in children". Arerugi 16 (3): 178–222.

Question 4.

A 45-year-old female complains of pain and swelling in both wrists and knees for three months. There is increased stiffness in the hands early in the morning, which lasts close to 40 minutes. On examination, the metacarpophalangeal joints and wrists are warm and tender. There are no other joint abnormalities. There is no alopecia, photosensitivity, kidney disease, or rash. What is the most likely diagnosis in this patient? (AIIMS)

1. Rheumatoid arthritis
2. Polymyalgia rheumatica
3. Gouty arthritis
4. Osteoarthritis

DISCUSSION:

Among articular disorders, gout and the spondyloarthropathies are more common in men, whereas rheumatoid arthritis and lupus are more frequent in women. This female patient has symmetrical arthritis with early morning stiffness and involvement of the wrist and metacarpophalangeal joints. All of them are characteristic features of rheumatoid arthritis. Osteoarthritis is seen in elderly patients and is non-inflammatory arthritis. Gout is common in males or postmenopausal females. Its most common presentation is acute mono-articular arthritis, frequently involving the first metatarsophalangeal joint. Swelling and tenderness of the metacarpophalangeal joints and wrists suggests articular disorder and hence rules out polymyalgia rheumatica.

ANSWER:

The correct answer is “rheumatoid arthritis.”

REFERENCES:

Maini R N. Chapter 19.5. Rheumatoid arthritis. In: Warrell DA, Cox TM, Firth JD, eds. Oxford

Textbook of Medicine. 5th edition.

Shah A, St. Clair E. Chapter 321. Rheumatoid Arthritis. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 5.

A 20-year-old woman presents with bilateral conductive deafness, palpable purpura on the legs, and hemoptysis. A radiograph of the chest shows a thin-walled cavity in the left lower zone. Investigations reveal red cell casts in the urine and an elevated serum creatinine level (3 mg/dL). What is the most probable diagnosis? (AI)

1. Henoch-Schonlein purpura
2. Polyarteritis nodosa
3. Granulomatosis with polyangiitis
4. Disseminated tuberculosis

DISCUSSION:

With vasculitis, lung involvement is commonly seen with granulomatosis with polyangiitis, microscopic PAN, Churg-Struass disease, and Takayasu's arteritis. Polyarteritis nodosa and Henoch-Schonlein purpura rarely show lung involvement. A triad of upper respiratory tract diseases (including rhinitis, sinusitis, and otitis media), lower respiratory disease (including pulmonary nodules, cavities, and hemoptysis) and glomerulonephritis (red cell cast, raised creatinine) characterizes granulomatosis with polyangiitis (Wegner's granulomatosis). Polyarteritis nodosa is not associated with glomerulonephritis (red cell casts), and Henoch-Schonlein purpura typically presents in children between 4 and 7 years of age. Disseminated tuberculosis is often associated with features such as fever, anorexia, and weight loss. In addition, a history of exposure to another patient with tuberculosis or a history of HIV infection is frequently elicited.

ANSWER:

The correct answer is "granulomatosis with polyangiitis."

REFERENCES:

Langford CA, Fauci AS. Chapter 326. The Vasculitis Syndromes. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Wells AU, M. du Bois R. Chapter 18.11.5. The lung in vasculitis. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 6.

A 34-year-old woman complains of paleness and bluish discoloration of the hands upon exposure to cold for the last five years and recent difficulty in swallowing solid food. On examination, there is tight skin over the face and fingers. Investigations show the presence of anti-centromere antibody in serum. What is the most likely cause? (AIIMS)

1. Systemic sclerosis
2. Mixed connective tissue disorder
3. SLE
4. Rheumatoid arthritis

DISCUSSION:

Raynaud's phenomenon is an episodic vasoconstriction in the fingers and toes that occurs in response to exposure to cold or emotional stress. It can be primary or secondary. Secondary Raynaud's is often due to associated connective tissue diseases or drugs such as atenolol. Dysphagia and sclerodactyly are two other clues, apart from Raynaud's phenomenon, that point toward the diagnosis of systemic sclerosis. It is a generalized connective tissue disorder causing diffuse fibrosis of the skin and affecting internal organs. It is divided into "diffuse cutaneous" and "limited cutaneous" systemic sclerosis. Mixed connective tissue disorder shows an overlap of clinical features of SLE, systemic sclerosis, and myositis. However, characteristic antibodies are anti-RNP (and not anti-centromere). Dysphagia and sclerodactyly are uncommon with SLE and rheumatoid arthritis.

ANSWER:

The correct answer is "systemic sclerosis."

REFERENCES:

Denton C P, Black C M. Chapter 19.11.3. Systemic sclerosis. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Varga J. Chapter 323. Systemic Sclerosis (Scleroderma) and Related Disorders. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 7.

A 60-year-old male present with left-sided headache and generalized aches and pains of three months' duration. For the last three days, he has had slight blurring of the vision from his left eye. The referring doctor has highlighted local tenderness over the left temporal artery and remarkably elevated erythrocyte sedimentation rate. Which of the following is the most likely diagnosis in this patient? (AIIMS)

1. Multiple myeloma
2. Disseminated carcinoma prostate
3. Paget's disease
4. Giant cell arteritis

DISCUSSION:

Symptoms associated with headache suggesting a serious underlying disorder include fever, neurological deficit, headache starting in old age, headache preceded by vomiting or associated with local tenderness over temporal artery, and sudden-onset very severe headache. Temporal arteritis, also known as giant cell arteritis (GCA), is an inflammatory disorder of elderly patients, mainly affecting the extra-cranial carotid circulation. It is characterized by headache, polymyalgia rheumatica (generalized aches and pains), and elevated ESR. Local tenderness over the temporal artery, when present, is an important clue to the diagnosis. The most serious complication of GCA is vision loss, though this can be prevented by prompt treatment with corticosteroids. Multiple myeloma is characterized by lytic bone lesions, hypercalcemia, and renal failure. Local tenderness over the temporal artery is unlikely. The first manifestation of Paget's disease is usually elevated alkaline phosphatase in the blood. Overall, the most common symptom is bone pain. Disseminated carcinoma of the prostate is associated with elevated levels of prostatic specific antigen. It is more likely to metastasize to the pelvic lymph nodes and the bones of the lumbar spine and pelvis.

ANSWER:
The correct answer is “giant cell arteritis.”

REFERENCES:
Langford CA, Fauci AS. Chapter 326. The Vasculitis Syndromes. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Gran JT. Chapter 19.11.4. Polymyalgia rheumatica and temporal arteritis. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 8.

A 23-year-old man presents with about a one-week history of foreign-body sensation in both eyes, painless swelling of the right ankle joint, bilateral heel pain, and painless ulcers on his penis. He was treated for nongonococcal urethritis about one week ago. Serum rheumatoid factor was negative. Which of the following is the most likely diagnosis in this case? (AIIMS)

1. Ankylosing spondylitis
2. Reactive arthritis
3. Acute gouty arthritis
4. SLE

DISCUSSION:
Seronegative spondyloarthritides mainly include ankylosing spondylitis, reactive arthritis, psoriatic arthritis, and enteropathic arthritis. They mainly affect young males and share certain features such as inflammatory arthritis of the spine and sacroiliac joints, asymmetric oligoarthritis of the large peripheral joints, enthesopathy, the absence of rheumatoid factor and ANA in the serum, and a striking association with HLA-B27. This patient shows characteristic lesion on the penis (circinate balanitis), evidence of previous urogenital infection, asymmetric oligoarthritis, and conjunctivitis, all suggesting reactive arthritis. Ankylosing spondylitis presents mainly with backache with variable peripheral joint involvement. SLE is found more commonly in females and has a malar rash, among other features. Acute gouty arthritis typically affects the first metatarsophalangeal joint without any skin or eye changes.

ANSWER:
The correct answer is “reactive arthritis.”

REFERENCES:
Braun J, Sieper J. Chapter 19.6. Ankylosing spondylitis, other spondyloarthritides, and related conditions. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Taurog JD. Chapter 325. The Spondyloarthritides. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 9.

A 12-year-old boy is brought to a clinic with a palpable purpuric rash over the buttocks and lower limbs. He also complains of pain in his knees and hands, and in the last six hours, he has developed abdominal pain. What is the most likely diagnosis? (AI)

1. Henoch-Schonlein Purpura
2. Sweet’s syndrome
3. Kawasaki disease
4. Hemochromatosis

DISCUSSION:
Sweet's syndrome (febrile neutrophilic dermatosis) is characterized by the sudden onset of fever, leukocytosis, and tender, erythematous, well-demarcated papules and plaques with neutrophilic infiltration. It is a reactive phenomenon and can be idiopathic, associated with malignancies, or drug-induced. Abdominal symptoms are not seen in Sweet’s syndrome. Kawasaki disease, also known as mucocutaneous lymph node syndrome, is characterized by the presence of fever, rash, conjunctivitis, and cervical lymphadenopathy. Peak incidence is seen below five years of age. Hemochromatosis is a result of iron overload and may present with cirrhosis of the liver, diabetes, cardiomyopathy, arthritis, and brown discoloration of the skin. Henoch-Schonlein purpura is typically associated with abdominal pain, palpable purpura, and arthritis. This disease is often preceded by an infection, such as a throat infection.

ANSWER:
The correct answer is “Henoch-Schonlein purpura.”

REFERENCES:
Langford CA, Fauci AS. Chapter 326. The Vasculitis Syndromes. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Barratt J, Feehally J. Chapter 21.8.1. Immunoglobulin A nephropathy and Henoch–Schönlein purpura. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 10.

A 32-year-old female presents wiyj polyarthralgias of two months’ duration. She has been having difficulty in swallowing solid food and frequent heartburn for the last year. Examination reveals a malar rash and skin stiffness over the face and fingers. She denies having taken any medicines prior to the onset of symptoms. There is proximal muscle weakness, and anti-RNP and ANA antibodies are present in serum. Which is the most likely diagnosis in this patient? (AIIMS)

1. Systemic lupus erythematosus
2. Mixed connective tissue disorder
3. Drug-induced lupus
4. Dermatomyositis

DISCUSSION:
Features favoring SLE are a woman of childbearing age having a malar rash, while those favoring systemic sclerosis are dysphagia and skin stiffness over the face and fingers. A feature favoring dermatomyositis is proximal muscle weakness. ANA can be positive in all the mentioned conditions. The most likely diagnosis in this case is mixed connective tissue disorder. This disorder often shows an overlap of clinical features of SLE, systemic sclerosis, and myositis. The characteristic antibody associated with mixed connective tissue disorder is anti-RNP. Dermatomyositis is characterized by a periorbital heliotrope rash and Gottron’s papules. Diagnosis of dermatomyositis is confirmed by muscle biopsy. Anti-histone antibody is typical of drug-induced lupus (which is unlikely in the absence of any history of drug intake prior to the onset of symptoms).

ANSWER:
The correct answer is “mixed connective tissue disorder.”

REFERENCES:
Denton C P, Black C M. Chapter 19.11.3. Systemic sclerosis. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Varga J. Chapter 323. Systemic Sclerosis (Scleroderma) and Related Disorders. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 11.

A 60-year-old female presents with two months’ history of a feeling of dry mouth and a gritty sensation in her eyes. The patient is on no medications. On exam, the buccal mucosa appears dry, and the salivary glands are enlarged bilaterally. Serum antinuclear antibodies are positive. What is the most likely diagnosis in this patient? (AIIMS)

1. SLE
2. Mixed connective tissue disorder
3. Sjögren’s syndrome
4. Behcet’s disease

DISCUSSION:
Antinuclear antibodies are present in many disorders, such as SLE, scleroderma, Sjögren’s syndrome, and rheumatoid arthritis, among others. Clinically, their repeated absence goes against the diagnosis of SLE. However, in other disorders, they have a limited diagnostic role. Features of dry eyes and dry mouth in a 60-year-old female suggest the diagnosis of Sjögren’s syndrome. Sjogren’s syndrome can be primary or secondary. Secondary Sjögren’s syndrome is associated with various disorders such as rheumatoid arthritis, SLE, mixed connective tissue disorder, and others. However, no additional clinical features are provided to consider an alternate diagnosis. Behcet’s disease involves oral and genital ulcers with uveitis.

ANSWER:
The correct answer is “Sjögren’s syndrome.”

REFERENCES:
Moutsopoulos HM, Tzioufas AG. Chapter 324. Sjögren's Syndrome. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Venables P J W. Chapter 19.11.6 Sjögren’s syndrome. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 12.

A 22-year-old male presents with the insidious onset of low back pain improved with exercise and worsened by rest. There is no history of diarrhea, conjunctivitis, urethritis, or nail changes. On examination, the patient has reduced lumbar flexion and extension. Serum RA factor and antinuclear antibodies are negative. Imaging reveals evidence of widening and sclerosis of the sacroiliac joints. What is the most likely diagnosis in this patient? (AIIMS)

1. Rheumatoid arthritis
2. Ankylosing spondylitis
3. Diffuse idiopathic skeletal hyperostosis
4. Reactive arthritis

DISCUSSION:
A young male presenting with insidious onset inflammatory back pain (worse at rest, better with activity) with limited spinal mobility raises the possibility of seronegative spondyloarthritides. They include ankylosing spondylitis, reactive arthritis, psoriatic arthritis, and enteropathic arthritis. Ankylosing spondylitis is the most common type of seronegative spondyloarthritides. Reactive arthritis may be associated with conjunctivitis and urethritis, while enteropathic arthritis is associated with bowel-related symptoms and psoriatic arthritis may be associated with nail changes. Rheumatoid arthritis spares the sacroiliac joints and only affects the cervical component of the spine. Diffuse idiopathic skeletal hyperostosis occurs in middle-aged and elderly patients and is often asymptomatic or associated with thoracic spinal pain.

ANSWER:
The correct answer is “ankylosing spondylitis.”

REFERENCES:
Braun J, Sieper J. Chapter 19.6. Ankylosing spondylitis, other spondyloarthritides, and related conditions. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Taurog JD. Chapter 325. The Spondyloarthritides. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 13.

A 35-year-old male presents with severe pain, swelling, and redness in the left big toe since early morning. There is no history of trauma. The patient is taking a diuretic for hypertension. Which one of the following is the most likely diagnosis in this patient? (PGI)

1. Rheumatoid arthritis
2. Gouty arthritis
3. Pseudogout
4. Septic arthritis

DISCUSSION:
Acute-onset arthritis involving a single joint raises the possibility of gout, pseudogout, septic arthritis, or reactive arthritis. Septic arthritis is often associated with trauma or infection of the surrounding area. Reactive arthritis is often preceded by infection of the gastrointestinal or urogenital system. Gout most frequently manifests as acute monoarticular arthritis, affecting the metatarsophalangeal joint first. It is more common in males. It is associated with increased serum uric acid level, and diuretics often worsen hyperuricemia. Rheumatoid arthritis is often symmetric polyarthritis. Pseudogout most frequently affects the knee joint and is most often seen in elderly patients.

ANSWER:
The correct answer is “gouty arthritis.”

REFERENCES:

Schumacher H, Chen LX. Chapter 333. Gout and Other Crystal-Associated Arthropathies. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Edward Roddy E, Doherty M. Chapter 19.10. Crystal-related arthropathies. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 14.

A 70-year-old male develops the sudden onset of severe left knee pain. The knee is red, swollen, and tender. He has a known case of hyperparathyroidism. An x-ray of the knee shows linear calcification. What is the most likely diagnosis in this patient? (AIIMS)

1. Pseudogout
2. Gout
3. Rheumatoid arthritis
4. Osteoarthritis

DISCUSSION:

Linear calcification or chondrocalcinosis refers to the radiographic evidence of calcification in the hyaline and/or fibrocartilage and is due to calcium pyrophosphate dihydrate crystal deposition. Chondrocalcinosis is often asymptomatic but can cause acute synovitis, which is known as pseudogout. Pseudogout is found mainly in elderly patients, and the most common joint to be affected is the knee joint. Definitive diagnosis requires demonstration of typical rhomboid or rod-like crystals in the synovial fluid or articular tissue. Hyperparathyroidism, hemochromatosis, and hypomagnesemia may be associated with pseudogout. Other mentioned conditions do not show linear calcification on X-ray. Gout is due to the deposition of urate crystals, and the most commonly affected joint is the first metatarsophalangeal joint. Osteoarthritis is a non-inflammatory disease. Rheumatoid arthritis presents as symmetric polyarthritis and is more common in females.

ANSWER:

The correct answer is “pseudogout.”

REFERENCES:
Schumacher H, Chen LX. Chapter 333. Gout and Other Crystal-Associated Arthropathies. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Edward Roddy E, Doherty M. Chapter 19.10. Crystal-related arthropathies. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 15.

A 30-year-old male patient presents with a four-month history of numbness in the right upper and both lower limbs. Examination reveals asymmetric neuropathy, high BP (160/110 mm Hg), normal peripheral pulses, and palpable purpura. Blood tests reveal the presence of HBsAg; however, antinuclear antibody, antibody to ds-DNA, and ANCA are negative. Arteriogram shows multiple aneurysms in the mesenteric and hepatic arteries. The most likely diagnosis is: (AIIMS)

1. Polyarteritis nodosa
2. Systemic lupus erythematosus
3. Wegener's granulomatosis
4. Microscopic polyangiitis

DISCUSSION:
Granulomatosis with polyangiitis (Wegner's), Churg-Strauss disease, and microscopic polyangiitis are also known as ANCA-associated vasculitis. Arteriogram (showing aneurysm) is useful in the diagnosis of certain vasculitic syndromes involving large and medium-sized vessels such as polyarteritis nodosa, isolated CNS vasculitis, and Takayasu's arteritis. Polyarteritis nodosa (PAN) is defined as a multisystem necrotizing inflammation of small to medium-sized muscular arteries, without glomerulonephritis or the involvement of venules. Mononeuritis multiplex (asymmetrical neuropathy) is seen in about half of patients with PAN. Renal involvement in PAN is in the form of arteritis without glomerulonephritis and manifests as hypertension and renal insufficiency. The presence of HBsAg is an important clue to the diagnosis of polyarteritis nodosa, although HBV accounts for less than 10% of all PAN cases. Arteriogram of a patient with polyarteritis nodosa characteristically shows sparing pulmonary arteries and the involvement of renal and other visceral arteries. There are no diagnostic serologic tests for PAN, and ANCA is negative. Antinuclear

antibody is a good screening test for SLE. Its absence makes the diagnosis of SLE unlikely. Similarly, ANCA negativity is a clue that ANCA-associated vasculitides are less likely.

ANSWER:
The correct answer is “polyarteritis nodosa.”

REFERENCES:
Langford CA, Fauci AS. Chapter 326. The Vasculitis Syndromes. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Jayne D. Chapter 21.10.2. The kidney in systematic vasculitis. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 16.

A 30-year-old female with rheumatoid arthritis of five years’ duration complains of pain in her right index and middle fingers over the past six weeks. The pain seems especially severe at night, often awakening her from sleep. The most likely cause of this patient's complaint is: (AIIMS)

1. Atlanto-axial subluxation of the cervical spine
2. Sensory peripheral neuropathy
3. Carpal tunnel syndrome
4. Rheumatoid vasculitis

DISCUSSION:
Peripheral nerve involvement can be in the form of mononeuritis simplex (a single peripheral nerve is involved), mononeuritis multiplex (involvement of more than one peripheral nerve in a non-contiguous manner), or polyneuropathy (involvement of multiple peripheral nerves in contiguous and symmetrical manner). Pain in the right index and middle fingers raises the possibility of either median nerve involvement or right C6–C7 root involvement. Nocturnal worsening of pain is characteristic of carpal tunnel syndrome. This is attributed to abnormal posture of the wrist while sleeping, leading to an increase in intra-carpal tunnel pressure. Atlanto-axial subluxation leads to cervical cord compression, which may lead to tetraparesis. Rheumatoid vasculitis is often associated with

mononeuritis multiplex. Sensory peripheral neuropathy would produce sensory loss in the “glove and stocking” distribution.

ANSWER:

The correct answer is “carpal tunnel syndrome.”

REFERENCES:

Maini R N. Chapter 19.5. Rheumatoid arthritis. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Shah A, St. Clair E. Chapter 321. Rheumatoid Arthritis. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

RHEUMATOLOGY SECTION TWO - VASCULITIC SYNDROME

Question 17.

All of the following except one are characteristic features of granulomatosis with polyangiitis (Wegner's granulomatosis). Which is the exception? (AIIMS)

1. Nodular pulmonary lesion
2. Intractable rhinitis and sinusitis
3. Terminal uremia
4. Congestive cardiac failure

DISCUSSION:

A triad of upper respiratory tract disease (including rhinitis, sinusitis, and otitis media), lower respiratory disease (including pulmonary nodules and cavities), and glomerulonephritis characterizes granulomatosis with polyangiitis (Wegner's granulomatosis). Renal disease is the major cause of mortality in this disease. The heart, gastrointestinal tract, and central nervous system are infrequently affected in this disorder. Congestive cardiac failure is not a typical feature of Wegner’s granulomatosis.

ANSWER:

The correct answer is “congestive heart failure.”

REFERENCE:

Seo P, Stone JH (July 2004). The antineutrophil cytoplasmic antibody-associated vasculitides. Am. J. Med. 117 (1): 39–50.

Question 18.

All of the following statements except one are true of polyarteritis nodosa (PAN). Which is the exception? (AI)

1. Associated with hepatitis B infection
2. Large-vessel vasculitis
3. Mononeuritis multiplex
4. Associated with hypertension

DISCUSSION:
Mononeuritis multiplex is seen in about half of patients with PAN. Renal involvement in PAN is in the form of arteritis without glomerulonephritis and manifests as hypertension and renal insufficiency. HBV was once the cause of up to 30% of PAN cases; however, its frequency has decreased over period of time and is now estimated to account for less than 10% of all PAN cases. Polyarteritis nodosa (PAN) is an example of medium-vessel (not large-vessel) vasculitis. Examples of large-vessel vasculitides are giant cell arteritis, Takayasu's arteritis, and isolated CNS angiitis.

ANSWER:
The correct answer is “large-vessel vasculitis.”

REFERENCE:
Stone JH. Polyarteritis nodosa. JAMA. Oct 2 2002;288(13):1632-9.

Question 19.

In granulomatosis with polyangiitis (Wegner's granulomatosis), the treatment of choice is: (PGI)

1. Splenectomy
2. Antibiotics
3. Cyclophosphamide
4. Conservative

DISCUSSION:

Cyclophosphamide and glucocorticoids are used as remission-induction therapy for severe antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis, including Wegner's granulomatosis. Without treatment, Wegner's granulomatosis can be rapidly fatal. After remission induction, remission maintenance is usually achieved either with azathioprine or with methotrexate. Adjunctive plasmapharesis is sometimes beneficial in patients with rapidly progressive glomerulonephritis.

ANSWER:
The correct answer is "cyclophosphamide."

REFERENCE:

Stone JH et al: Rituximab versus cyclophosphamide for ANCA-associated vasculitis. N Engl J Med 363:221, 2010.

Question 20.

Henoch-Schonlein purpura is characterized by the deposition of the following immunoglobulin around the vessels: (AIIMS)

1. IgM
2. IgG
3. IgA
4. IgE

DISCUSSION:
Henoch–Schönlein purpura is a small-vessel vasculitis in which complexes of IgA and C3 are deposited on arterioles, capillaries, and venules. The serum levels of IgA are elevated in about 50% of patients with HSP. Renal biopsy findings are similar to those in IgA nephropathy.

ANSWER:
The correct answer is "IgA."

REFERENCE:

Saulsbury FT (2001). "Henoch-Schönlein purpura". Current Opinion in Rheumatology 13 (1): 35–40.

Question 21.

All of the following statements except one are true of polyarteritis nodosa (PAN). Which is the exception? (UPSC)

1. It is a multi-system necrotizing vasculitis.
2. Small and medium-sized vessels are involved.
3. Pulmonary artery involvement is a characteristic feature.
4. Hepatitis B virus may play a causative role.

DISCUSSION:

Polyarteritis nodosa (PAN) is defined as a multisystem necrotizing inflammation of small to medium-sized muscular arteries, without glomerulonephritis or involvement of venules. Hepatitis B virus-associated polyarteritis nodosa is a typical form of classic PAN whose pathogenesis has been attributed to immune-complex deposition. Among vasculitis, lung involvement is commonly seen with granulomatosis with polyangiitis, microscopic PAN, Churg-Struass disease, and Takayasu's arteritis. Polyarteritis nodosa characteristically involves renal and other visceral arteries and spares pulmonary arteries.

ANSWER:

The correct answer is "pulmonary artery involvement is a characteristic feature."

REFERENCE:

Colmegna I, Maldonado-Cocco JA. Polyarteritis nodosa revisited. Curr Rheumatol Rep. Aug 2005;7(4):288-96.

Question 22.

All of the following statements except one are correct about giant cell arteritis. Identify the exception. (AI)

1. High-dose steroid is the drug of choice.
2. ESR is usually raised.
3. The renal artery is the most commonly involved vessel.
4. It mainly affects people above 60 years of age.

DISCUSSION:

Giant cell arteritis (GCA) is seen in elderly patients and presents with headache and raised ESR. The most serious complication of GCA is permanent blindness, though this can be prevented by prompt treatment with corticosteroids. Up to one-third of patients can have large-vessel disease manifesting as arm claudication (subclavian artery stenosis) or thoracic (and less commonly abdominal) aortic aneurysms. The temporal artery is the most commonly affected vessel; hence, giant cell arteritis is also known as temporal arteritis.

ANSWER:

The correct answer is, “The renal artery is the most commonly involved vessel.”

REFERENCE:

Salvarani C, et al. (2002). Polymyalgia rheumatica and giant-cell arteritis. N Engl J Med, **347**, 261–71.

Question 23.

In a patient of polyarteritis nodosa, arteriography may show aneurysm in arteries of all except one of the following organs. Which is the exception? (AIIMS)

1. Kidney
2. Lung
3. Liver
4. Pancreas

DISCUSSION:
A diagnosis of vasculitic syndrome is best secured by biopsy. Arteriogram (showing aneurysm) is useful in the diagnosis of certain vasculitic syndromes involving large and medium-sized vessels, such as polyarteritis nodosa, isolated CNS vasculitis, and Takayasu's arteritis. Polyarteritis nodosa characteristically spares pulmonary arteries and involves renal and other visceral arteries.

ANSWER:
The correct answer is "lung."

REFERENCE:
Stanson AW, Friese JL, Johnson CM, et al. Polyarteritis nodosa: spectrum of angiographic findings. Radiographics. Jan-Feb 2001;21(1):151-9.

Question 24.

In which of the following vasculitis syndrome is bronchial asthma a common presentation? (UPSC)

1. Wegener's granulomatosis
2. Microscopic polyarteritis
3. Polyarteritis nodosa
4. Churg-Strauss vasculitis

DISCUSSION:
Churg-Strauss syndrome (also known as allergic angiitis and granulomatosis) is an example of small-vessel vasculitis, which is frequently associated with ANCA. It usually manifests in three stages: the early (prodromal) stage with asthma and/or allergic rhinitis, the second stage with eosinophilia, and the third and final stage with vasculitis.

ANSWER:
The correct answer is "Churg-Strauss vasculitis."

REFERENCE:
Della Rossa A, Baldini C, Tavoni A, et al. (November 2002). "Churg-Strauss syndrome: clinical and serological features of 19 patients from a single Italian centre". Rheumatology (Oxford) 41 (11): 1286–94.

Question 25.

All of the following except one are the clinical manifestations of Kawasaki disease. Which is the exception? (AIIMS)

1. Coronary aneurysm
2. Cervical lymphadenopathy
3. Strawberry tongue
4. Exudative conjunctivitis

DISCUSSION:
Kawasaki disease, also known as mucocutaneous lymph node syndrome, is characterized by the presence of fever, rash, and cervical lymphadenopathy. It is a type of vasculitis, affecting mainly the medium-sized vessels. Coronary artery aneurysms are important complications of this disease, which are mainly seen in untreated children.

Bilateral conjunctival injection is one of the most common and characteristic findings of Kawasaki disease. It is mostly confined to the bulbar region and is non-exudative in nature.

ANSWER:
The correct answer is “exudative conjunctivitis.”

REFERENCE:
Ohno S, Miyajima T, Higuchi M, et al. Ocular manifestations of Kawasaki's disease (mucocutaneous lymph node syndrome). Am J Ophthalmol. Jun 1982;93(6):713-7.

Question 26.

Feature(s) of microscopic polyangiitis is/are: (PGI)

1. IgG deposits in the kidney
2. Bronchospasm
3. Renal involvement in 80% of cases
4. All of the above

DISCUSSION:

Bronchospasm is a feature of Churg-Strauss syndrome and not microscopic polyangiitis. Evidence of immune complex deposition is not found in microscopic polyangiitis. Microscopic polyangiitis is a systemic, pauci-immune, necrotizing, small-vessel vasculitis. Glomerulonephritis is very common (unlike classical PAN) in microscopic polyangiitis.

ANSWER:

The correct answer is “renal involvement in 80% of cases.”

REFERENCE:

Berden AE, et al. (2010). Histopathologic classification of ANCA-associated glomerulonephritis. J Am Soc Nephrol, 21, 1628–36.

Question 27.

All of the following except one are features of classical polyarteritis nodosa. Which is the exception? (PGI)

1. Hypertension
2. Glomerulonephritis
3. Mononeuritis multiplex
4. Testicular pain

DISCUSSION:

Polyarteritis nodosa (PAN) is an example of medium-vessel vasculitis. Mononeuritis multiplex is seen in about half of patients with PAN. Testicular pain can be seen in patients with polyarteritis nodosa, which is attributed to the vasculitis of testicular artery. Renal involvement in PAN is in the form of arteritis without glomerulonephritis and manifests as hypertension and renal insufficiency.

ANSWER:
The correct answer is “glomerulonephritis.”

REFERENCE:
Colmegna I, Maldonado-Cocco JA. Polyarteritis nodosa revisited. Curr Rheumatol Rep. Aug 2005;7(4):288-96.

Question 28.

All of the following features except one may be used to distinguish polyarteritis nodosa (PAN) from microscopic polyangiitis. Which is the exception? (PGI)

1. ANCA positivity
2. RBC cast in urine
3. Necrotizing vasculitis
4. HBV infection

DISCUSSION:
RBC cast in urine is suggestive of glomerulonephritis, which is seen in microscopic polyangiitis but not in PAN. ANCA positivity is seen with microscopic polyangiitis, but not with PAN. Hepatitis B infection is associated with PAN but not with microscopic polyangiitis. Necrotizing vasculitis is seen in both PAN and microscopic polyangiitis. However, there is a difference in the types of vessels involved. PAN affects small and medium-sized muscular arteries but not venules. Microscopic polyangiitis, on the other hand, affects capillaries, venules, or arterioles.

ANSWER:
The correct answer is “necrotizing vasculitis.”

REFERENCE:
Seo P, Stone JH (July 2004). 'The antineutrophil cytoplasmic antibody-associated vasculitides." Am. J. Med. 117 (1): 39–50.

Question 29.

Which of the following is the most frequent presenting symptom in patients with giant cell arteritis? (AI)

1. Headache
2. Jaw claudication
3. Diminished radial pulse
4. Blindness

DISCUSSION:
The most serious complication of temporal arteritis is permanent blindness, which can be prevented by prompt treatment with corticosteroids. Up to one-third of patients can have large-vessel disease manifesting as arm claudication (subclavian artery stenosis and diminished radial pulses) or thoracic (and less commonly abdominal) aortic aneurysms. Jaw claudication or pain while chewing is occasionally noted. However, the most common presentation is with headache and raised ESR.

ANSWER:
The correct answer is "headache."

REFERENCE:
Myklebust G, Gran JT (1996). A prospective study of 287 patients with polymyalgia rheumatica and temporal arteritis: clinical and laboratory manifestations at onset of disease and at the time of diagnosis. *Br J Rheumatol*, **35**, 1161–8.

Question 30.

All of the following features except one may be used to distinguish granulomatosis with polyangiitis from microscopic polyangiitis. Which is the exception? (PGI)

1. Granulomatous lesion
2. Upper airway disease
3. Pulmonary nodules
4. Renal lesion

DISCUSSION:

Granulomatous lesions, upper airway disease, and pulmonary nodules are seen in granulomatosis with polyangiitis, but not in microscopic polyangiitis. However, renal lesions are similar in both conditions: evidence of immune complex deposition is not found, and patients are treated with cyclophosphamide.

ANSWER:

The correct answer is “renal lesion.”

REFERENCE:

de Lind van Wijngaarden RA, *et al*. (2006). Clinical and histologic determinants of renal outcome in ANCA-associated vasculitis: a prospective analysis of 100 patients with severe renal involvement. *J Am Soc Nephrol*, **17**, 2264–74.

Question 31.

Which of the following is considered the best investigation for diagnosing giant cell arteritis? (AI)

1. Temporal artery biopsy
2. Color Doppler of the temporal artery
3. PET scan
4. MRA

DISCUSSION:
Giant cell arteritis presents with clinical features such as headache, high ESR, fever, girdle muscle stiffness, and pain (polymyalgia rheumatic) in an elderly patient. The best way to confirm the diagnosis is by performing a temporal artery biopsy. Typical findings on biopsy include nodular intimal thickening and medial granulomatous inflammation causing the fragmentation of internal elastic lamina. Ultrasonography, PET scan, and MRA are used with variable success rates. However, their sensitivity and specificity are inferior to those of temporal artery biopsy.

ANSWER:
The correct answer is "temporal artery biopsy."

REFERENCE:
Narvaez J, Bernad B, Roig-Vilaseca D, et al. Influence of previous corticosteroid therapy on temporal artery biopsy yield in giant cell arteritis. Semin Arthritis Rheum. Aug 2007;37(1):13-9.

Question 32.

All of the following except one are features of Kawasaki disease. Which is the exception? (AI, AIIMS)

1. Peak incidence at > 25 years of age
2. Aneurysm of coronary artery
3. Enlarged lymph nodes
4. Fever

DISCUSSION:
Kawasaki disease, also known as mucocutaneous lymph node syndrome, is characterized by the presence of fever, rash (which is never bullous or vesicular), non-purulent conjunctivitis, and cervical lymphadenopathy. It is a type of vasculitis affecting mainly the medium-sized vessels. Coronary artery aneurysms are important complications of this disease, mainly seen in untreated children. It is largely seen in children less than five years of age.

ANSWER:
The correct answer is “peak incidence at > 25 years of age.”

REFERENCE:
Falcini F: Kawasaki disease. Curr Opin Rheumatol 18:33, 2006.

Question 33.

All of the following except one are features of temporal arteritis. Which is the exception? (AI, AIIMS)

1. Polymyalgia rheumatica
2. Anemia
3. Low ESR
4. Sudden blindness

DISCUSSION:
Giant cell arteritis is mainly a disease of the elderly population and is frequently associated with fever, anemia, and polymyalgia rheumatica. The most dreaded complication is potential blindness, which can be prevented with the timely introduction of steroids. ESR is characteristically high in both temporal arteritis and polymyalgia rheumatica. Other causes of high ESR include pregnancy, inflammation, or anemia. Low ESR is observed in conditions such as polycythemia, sickle cell anemia, hereditary spherocytosis, and congestive heart failure.

ANSWER:
The correct answer is “low ESR.”

REFERENCE:
Myklebust G, Gran JT (1996). A prospective study of 287 patients with polymyalgia rheumatica and temporal arteritis: clinical and laboratory manifestations at onset of disease and at the time of diagnosis. *Br J Rheumatol*, **35**, 1161–8.

Question 34.

While evaluating a patient with suspected Henoch-Schönlein purpura, which of the following investigations is least useful? (PGI)

1. Urine routine and microscopy
2. Complete blood count
3. Chest X-ray
4. Skin biopsy

DISCUSSION:

Purpura, arthritis, and abdominal pain are known as the "classic triad" of Henoch-Schönlein purpura. The purpura typically appears on the legs and buttocks. About 20–40% of patients develop kidney involvement. Almost all have evidence of gross or microscopic hematuria and, less frequently, proteinuria on urinalysis. The complete blood count may reveal normal or raised platelet count and distinguishes it from diseases such as thrombotic thrombocytopenic purpura. Skin biopsy is useful to confirm the diagnosis, showing IgA and C3 deposition by immunofluorescence. Chest X-ray is the least useful in evaluating these patients.

ANSWER:

The correct answer is “chest X-ray.”

REFERENCE:

Saulsbury FT (1999). "Henoch-Schönlein purpura in children. Report of 100 patients and review of the literature". Medicine (Baltimore) **78** (6): 395–409.

Question 35.

A 60-year-old male presents with a left-sided headache and generalized aches and pains of three months’ duration. For the last three days, he has had slight blurring of the vision from his left eye. There is local tenderness over the left temporal artery and an elevated ESR. Which of the following is the most important intervention in this patient? (AIIMS)

1. Serial ESR measurement

2. Temporal artery biopsy
3. Giving 400 mg ibuprofen
4. Giving 60 mg prednisolone

DISCUSSION:
This patient is suffering from temporal arteritis. It is seen in elderly patients and presents with headache and raised ESR. Diagnosis is confirmed by performing temporal artery biopsy. The most serious complication of temporal arteritis is permanent blindness, which can be prevented with prompt treatment with corticosteroids. This clearly takes precedence over all other interventions.

ANSWER:
The correct answer is "giving 60 mg prednisolone."

REFERENCE:
Hayreh SS, Zimmerman B, Kardon RH (2002). Visual improvement with corticosteroid therapy in giant cell arteritis. Report of a large study and review of the literature. Acta Ophthalomol Scand, 80, 353–67.

Question 36.
The treatment of choice for Kawasaki disease is: (AIIMS)

1. Cyclosporine
2. Prednisolone
3. Immunoglobulins
4. Methotrexate

DISCUSSION:
Coronary artery aneurysms are important complications of Kawasaki disease, mainly seen in untreated children. Treatment should be started as soon as the diagnosis is made to prevent damage to the coronary arteries. Intravenous immunoglobulin together with aspirin is the standard treatment for Kawasaki disease. Affected children are advised to be vaccinated against varicella and influenza to

prevent Reye's syndrome caused by aspirin.

ANSWER:
The correct answer is “immunoglobulins.”

REFERENCE:
Gedalia A: Kawasaki disease: 40 years after the original report. Curr Rheumatol Rep 9:336, 2007.

Question 37.

Which one of the following features best describes polymyalgia rheumatica? (PGI)

1. A normal erythrocyte sedimentation rate
2. Oral steroids contraindicated
3. Stiffness and tenderness of girdle muscles
4. Mainly seen in patients below 40 years

DISCUSSION:
Polymyalgia rheumatica is seen in the elderly population. The erythrocyte sedimentation rate is frequently elevated. It is associated with temporal arteritis in about 15% of patients. Like temporal arteritis, there is dramatic response to oral steroids, but unlike temporal arteritis (which typically requires a higher dose), a low dose of steroids is usually sufficient to control symptoms. Polymyalgia rheumatica is characterized by generalized stiffness and muscle tenderness.

ANSWER:
The correct answer is “stiffness and tenderness of girdle muscles.”

REFERENCE:
Myklebust G, Gran JT (1996). A prospective study of 287 patients with polymyalgia rheumatica and temporal arteritis: clinical and laboratory manifestations at onset of disease and at the time of diagnosis. Br J Rheumatol, 35, 1161–8.

Question 38.

All of the following except one are the features of Kawasaki's disease. Which is the exception? (AI)

1. Coronary artery aneurysm
2. Conjunctival suffusion
3. Thrombocytopenia
4. Desquamation of the skin of the fingers

DISCUSSION:

Tomisaku Kawasaki in Japan first described Kawasaki's disease in 1967. The acronym FEBRILE is used to remember the clinical features of this disease: fever, exanthem, bulbar conjunctivitis, rash, internal organ involvement (coronary arteritis and aneurysm), lymphadenopathy, and extremity changes. During the subacute stage, thrombocytosis is usually seen. The platelet count begins to rise in the second week and continues to rise during the third week.

ANSWER:

The correct answer is "thrombocytopenia."

REFERENCE:

Kawasaki T (1967). "Acute febrile mucocutaneous syndrome with lymphoid involvement with specific desquamation of the fingers and toes in children". Arerugi 16 (3): 178–222.

Question 39.

All of the following features except one are seen in Henoch-Schonlein purpura (HSP). Which is the exception? (PGI)

1. Arthralgia
2. Glomerulonephritis
3. Raised serum IgA
4. Thrombocytopenia

DISCUSSION:

Henoch-Schonlein purpura is a common systemic vasculitis in children, but it occurs in adults as well. It is typically associated with abdominal pain, palpable purpura, and arthritis. Serum IgA level is elevated in about one-half of patients with HSP. Renal involvement is characterized by mild glomerulonephritis, associated with hematuria and proteinuria. Purpura in HSP is due to vasculitis and not thrombocytopenia.

ANSWER:

The correct answer is “thrombocytopenia.”

REFERENCE:

Davin JC, Ten Berge IJ, Weening JJ (2001). What is the difference between IgA nephropathy and Henoch–Schönlein purpura nephritis? Kidney Int, 59, 823–34.

RHEUMATOLOGY SECTION THREE - INFLAMMATORY ARTHRITIS

Question 40.

Which of the following is least likely to be associated with gout? (JIPMER)

1. Thiazide diuretics
2. Rheumatoid arthritis
3. Low dose aspirin
4. Osteoarthritis

DISCUSSION:

Gout is associated with osteoarthritis, Heberden's and Bouchard's nodes being common sites of involvement. Drugs frequently implicated in development of gout include aspirin, thiazide diuretics, cyclosporin and pyrazinamide.

Typically, patients with rheumatoid arthritis do not develop gout. The reason for this strong negative association is not known.

ANSWER:

The correct answer is 'rheumatoid arthritis'.

REFERENCES:

Edward Roddy E, Doherty M. Chapter 19.10. Crystal-related arthropathies. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Roddy E, Doherty M. Chapter 19.10. Crystal-related arthropathies. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 41.

All of the following except one are joints frequently involved in ankylosing spondylitis. Which is the exception? (AI)

1. Small joints of hands
2. Sacroiliac joints
3. Spine
4. Hip joints

DISCUSSION:

Ankylosing spondylitis is chronic inflammatory arthritis predominantly affecting the sacroiliac joints and spine. Peripheral joints involved are mainly knees, ankles, hips, and shoulders. The most common indication for surgery in patients with ankylosing spondylitis is severe hip joint arthritis, and development of hip arthritis within the first 2 years of disease predicts poor outcome.

Small joints are rarely affected.

ANSWER:

The correct answer is ‘small joints of hands’.

REFERENCE:

Braun J, Sieper J (2007). Ankylosing spondylitis. Lancet 369, 1379–90.

Question 42.

Which of the following is the commonest cardiac lesion in rheumatoid arthritis? (PGI)

1. Pancarditis
2. Pericarditis
3. Myocarditis
4. Endocarditis

DISCUSSION:

The commonest cardiac manifestation in rheumatoid arthritis is pericarditis, affecting approximately 10% of patients. Rarely, symptomatic pericardial effusions and constrictive pericarditis have been described. Myocardial involvement in RA can be in the form of heart muscle containing rheumatoid nodules or infiltrated with amyloid. Mitral valve incompetence is the most common valvular abnormality.

ANSWER:

The correct answer is 'pericarditis'.

REFERENCES:

Maini R N. Chapter 19.5. Rheumatoid arthritis. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Shah A, St. Clair E. Chapter 321. Rheumatoid Arthritis. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 43.

In a patient with gouty arthritis, which of the following is most useful in establishing the diagnosis? (AIIMS)

1. Serum uric acid
2. Uric acid in urine
3. Urate crystal in synovial fluid
4. Serum calcium level

DISCUSSION:

Gouty arthritis is associated with a chronic raised level of serum uric acid. However, the majority of patients with hyperuricemia are asymptomatic, so high serum uric acid level is not enough to make a diagnosis of gout. Measurement of uric acid in urine (24 hours' uric acid in urine) helps to understand the underlying defect (underexcretors or overproducers) of gout. Serum calcium has no role in diagnosis of gouty arthritis.

To establish diagnosis of gouty arthritis, demonstration of urate crystals in synovial fluid is required.

ANSWER:
The correct answer is ‘urate crystals in synovial fluid’.

REFERENCES:
Schumacher H, Chen LX. Chapter 333. Gout and Other Crystal-Associated Arthropathies. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Schlesinger N (March 2010). Diagnosing and treating gout: a review to aid primary care physicians. Postgrad Med 122 (2): 157–61.

Question 44.

Which one of the following statements is correct about ankylosing spondylitis? (PGI)

1. Peripheral joint is rarely involved
2. Sacroiliac joints are usually spared
3. May be associated with anterior uveitis
4. More common in females

DISCUSSION:
Ankylosing spondylitis is an inflammatory disorder that primarily affects the axial skeleton, often starting with the sacroiliac joint. The age of onset is commonly in the twenties, and males are affected more commonly than females. Transient or persistent peripheral arthritis is frequently seen in these patients.

Acute anterior uveitis is the most common extra-articular manifestation (seen in about 40% of patients) and can occur before the joint symptoms.

ANSWER:
The correct answer is ‘may be associated with anterior uveitis’.

REFERENCE:

Gran JT, Skomsvoll JF (1997). The outcome of ankylosing spondylitis: a study of 100 patients. Br J Rheumatol 36, 766–71.

Question 45.

All of the following statements except one are correct about gout. Which is the exception? (PGI)

1. Occurs due to accumulation of urate crystals in joint
2. Can be precipitated by pyrazinamide
3. Occurs more frequently in females
4. Hyperuricemia in primary gout is mainly due to decreased excretion of uric acid

DISCUSSION:

Gout is a metabolic disease associated with hyperuricemia and urate crystal deposition in joints. In patients with primary gout, the hyperuricemia is mainly due to reduced excretion of uric acid. The next common cause is increased production. Drugs frequently implicated in development of gout include aspirin, thiazide diuretics, cyclosporin, pyrazinamide and niacin.

Gout is more frequent in males.

ANSWER:

The correct answer is ‘occurs more frequently in females’.

REFERENCES:

Edward Roddy E, Doherty M. Chapter 19.10. Crystal-related arthropathies. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Choi HK, Mount DB, Reginato AM (2005). Pathogenesis of gout. Ann Intern Med 143, 499–516.

Question 46.

Which one of the following is least likely to occur as a part of seropositive rheumatoid arthritis? (UPSC)

1. Splenomegaly
2. Pericarditis
3. Pleural effusion
4. Hepatomegaly

DISCUSSION:

Pericarditis and pleural effusion occur as part of serositis. Splenomegaly is described as part of Felty's syndrome (triad of splenomegaly, neutropenia and rheumatoid arthritis).

Hepatomegaly is not typically seen in rheumatoid arthritis. Rarely, in Felty's syndrome, Kupffer cell activation by cytokines is associated with nodular hyperplasia of the liver, which may be palpably enlarged.

ANSWER:

The correct answer is 'hepatomegaly'.

REFERENCES:

Maini R N. Chapter 19.5. Rheumatoid arthritis. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Turesson C, O'Fallon WM, Crowson CS, Gabriel SE, Matteson EL (2003). Extra-articular disease manifestations in rheumatoid arthritis: incidence trends and risk factors over 46 years. Ann Rheum Dis 62 (8): 722–7.

Question 47.

Pseudogout can be distinguished from gout by means of: (PGI)

1. Acute onset

2. Positive birefringent crystals
3. Association with diabetes mellitus
4. Involvement of large joints

DISCUSSION:
Both gout and pseudogout can be acute in onset, may involve large joints and may be associated with diabetes.

Pseudogout is found mainly in elderly patients and the most common joint to be affected is the knee joint. Definitive diagnosis requires demonstration of typical rhomboid or rod-like crystals in synovial fluid or articular tissue. Pseudogout crystals show a weakly positive birefringence, whereas monosodium urate crystals of gout show a strongly negative birefringence.

ANSWER:
The correct answer is ‘positive birefringent crystals’.

REFERENCES:
Schumacher H, Chen LX. Chapter 333. Gout and Other Crystal-Associated Arthropathies. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Dieppe P, Swan A (1 May 1999). Identification of crystals in synovial fluid. Ann Rheum Dis 58 (5): 261–3.

Question 48.
Which of the following joints is characteristically involved in rheumatoid arthritis? (AIIMS)

1. Spine
2. Knee joint
3. Metacarpophalangeal joint
4. Distal interphalangeal joint

DISCUSSION:
The typical joints involved in rheumatoid arthritis are the metacarpophalangeal and proximal interphalangeal joints. Other frequently involved sites include the wrists, shoulders, knees, elbow joints, atlantoaxial joints of the cervical spine, and hip joints.

Distal interphalangeal joints involvement is more suggestive of other conditions such as osteoarthritis or psoriatic arthritis.

ANSWER:
The correct answer is ‘metacarpophalangeal joint’.

REFERENCES:
Shah A, St. Clair E. Chapter 321. Rheumatoid Arthritis. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Huizinga TW, et al. (6 July 2010). In the clinic. Rheumatoid arthritis. Ann Intern Med 153 (1): ITC1–15.

Question 49.

Pseudogout has been associated with or encountered in: (PGI)

1. Hypomagnesemia
2. Hemochromatosis
3. Hyperparathyroidism
4. All of the above

DISCUSSION:
Pseudogout is due to deposition of calcium pyrophosphate dihydrate (CPPD) crystals in articular tissues. It is most common in elderly patients, and, in most cases, is asymptomatic. The majority of cases are idiopathic. A minority of patients with CPPD arthropathy have metabolic abnormalities (such as hyperparathyroidism, hemochromatosis, and hypomagnesemia) or hereditary CPPD disease.

ANSWER:
The correct answer is ‘all of the above’.

REFERENCES:
Edward Roddy E, Doherty M. Chapter 19.10. Crystal-related arthropathies. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Wright GD, Doherty M (1997). Calcium pyrophosphate crystal deposition is not always “wear and tear” or aging. Ann Rheum Dis 56 (10): 586–8.

Question 50.

Which of the following is true of psoriatic arthritis? (JIPMER)

1. Involves distal joints of hand
2. Pencil-in-cup deformity may be seen
3. Sacroiliitis may be seen
4. All are correct

DISCUSSION:
Psoriatic arthritis is a type of seronegative spondyloarthritides (RA negative and HLA-B27 positive), which almost always accompanies skin disease, especially nail changes. Like other seronegative spondyloarthritides, it shows sacroiliitis. In addition, it commonly affects distal interphalangeal joints (as does osteoarthritis). Classical deformity, as seen on X-ray, is called ‘pencil-in-cup deformity’ or ‘cup-and-pencil deformity’.

ANSWER:
The correct answer is ‘all are correct’.

REFERENCE:
Braun J, Sieper J. Chapter 19.6. Ankylosing spondylitis, other spondyloarthritides, and related conditions. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 51.

Which of the following joints is most frequently involved in a patient with an acute attack of gout? (AIIMS)

1. Knee joint
2. Ankle joint
3. 1st metatarsophalangeal joint
4. 1st metacarpophalangeal joint

DISCUSSION:

Acute monoarticular arthritis is the most common early clinical manifestation of gout. In more than half of affected patients, the first metatarsophalangeal joint (podagra) is involved. Other common sites are the midtarsal joints, ankle, knee, wrist, and elbow joints. Inflamed Heberden's or Bouchard's nodes may also be the first manifestation of gouty arthritis. Hips and shoulders are rarely affected.

ANSWER:

The correct answer is '1st metatarsophalangeal joint'.

REFERENCES:

Schumacher H, Chen LX. Chapter 333. Gout and Other Crystal-Associated Arthropathies. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Chen LX, Schumacher HR (October 2008). Gout: an evidence-based review. J Clin Rheumatol 14 (5 Suppl): S55–62.

Question 52.

All of the following except one are features of rheumatoid arthritis. Which is the exception? (AI)

1. Anti IgG antibody
2. Antinuclear antibody
3. Early morning stiffness
4. No joint deformity

DISCUSSION:
IgM, IgG, and IgA isotypes of RA factor occur in blood (IgM isotype is most commonly measured). Antinuclear antibody, rheumatoid factor, and anti-CCP antibodies are frequently seen in rheumatoid arthritis. Early morning stiffness lasting more than one hour is common in this condition.

Joint deformities are common with rheumatoid arthritis. Characteristic joint deformities include 'swan neck' deformity, the ‘boutonnière’ deformity, and the ‘Z-line’ deformity of the thumb.

ANSWER:
The correct answer is ‘no joint deformity’.

REFERENCES:
Maini R N. Chapter 19.5. Rheumatoid arthritis. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Majithia V, Geraci SA (2007). Rheumatoid arthritis: diagnosis and management. Am J Med 120 (11): 936–9.

Question 53.

Rheumatoid arthritis commonly affects which one of the following regions of the spine? (AIIMS)

1. Cervical spine
2. Lumbar spine
3. Thoracic spine
4. Sacral spine

DISCUSSION:
Rheumatoid arthritis affects the cervical spine, and, unlike ankylosing spondylitis, does not usually affect the thoracic and lumbosacral spine. This is important to know, as rheumatoid arthritis can potentially cause atlanto-axial subluxation and consequent compressive myelopathy.

ANSWER:

The correct answer is ‘cervical spine’.

REFERENCES:

Shah A, St. Clair E. Chapter 321. Rheumatoid Arthritis. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Mathews JA (May 1969). Atlanto-axial subluxation in rheumatoid arthritis. Ann Rheum Dis 28 (3): 260-6.

Question 54.

Prolonged allopurinol therapy in a patient with gout is indicated in all except one of the following conditions. Which is the exception? (UPSC)

1. Acute gouty arthritis
2. Tophi
3. Urate nephropathy
4. Evidence of bone /joint damage

DISCUSSION:

Allopurinol should not be started during an acute attack, as it can result in sudden lowering of uric acid. Sudden changes in concentration may lead to further attacks of gouty arthritis.

Hypouricemic therapy in the form of allopurinol is usually indicated when there have been two or more acute attacks of gout, when gout is associated with tophi, bone or joint damage, high serum uric acid level (>9.0 mg/dL), or there are uric acid stones.

ANSWER:

The correct answer is ‘acute gouty arthritis’.

REFERENCES:

Edward Roddy E, Doherty M. Chapter 19.10. Crystal-related arthropathies. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Chen LX, Schumacher HR (October 2008). Gout: an evidence-based review. J Clin Rheumatol 14 (5 Suppl): S55–62.

Question 55.

Which of the following is true regarding rheumatoid arthritis? (AI)

1. Typically spares the cervical spine
2. Causes pleural effusion with low sugar
3. Pulmonary nodules are absent
4. Enthesitis is prominent

DISCUSSION:

The typical joint involvement seen in rheumatoid arthritis includes metacarpophalangeal and proximal interphalangeal joints. Rheumatoid arthritis affects the cervical spine, and, unlike ankylosing spondylitis, spares the thoracic and lumbosacral spine. Pulmonary nodules and subcutaneous nodules are frequently observed. Enthesitis (inflammation at tendon, ligament, or joint capsule insertions) is a characteristic feature of spondyloarthritides, and not rheumatoid arthritis.

Empyema and rheumatoid arthritis are two important causes of pleural effusion with low sugar.

ANSWER:

The correct answer is ‘causes pleural effusion with low sugar’.

REFERENCES:

Shah A, St. Clair E. Chapter 321. Rheumatoid Arthritis. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

McGonagle DI, Khan MA, Marzo-Ortega H, O'Connor P, Gibbon W, Emery P (July 1999). Enthesitis in spondyloarthropathy. Curr Opin Rheumatol (4): 244–50.

Question 56.

All of the following except one are characteristic features of ankylosing spondylitis. Which is the exception? (AI)

1. Strongly positive rheumatoid factor
2. Association with HLA-B27
3. Anterior uveitis
4. 'Bamboo' spine on X-ray

DISCUSSION:

Ankylosing spondylitis, or Marie-Strümpell disease, is a chronic inflammatory disease of the axial skeleton with involvement of various peripheral joints and extra-articular features. It shows HLA-B27 positivity in the majority of patients, and anterior uveitis is the most common extra-articular manifestation. A bamboo spine refers to the vertebral body fusion by marginal syndesmophytes in patients with ankylosing spondylitis.

Ankylosing spondylitis is a type of seronegative spondyloarthritides. Here, rheumatoid factor and antinuclear antibodies are absent.

ANSWER:

The correct answer is 'strongly positive rheumatoid factor'.

REFERENCE:

Sieper J, Braun J, Rudwaleit M, Boonen A, Zink A (2002). Ankylosing spondylitis: an overview. Ann Rheum Dis 61 (3): iii8.

Question 57.

All of the following except one can be used to prevent attacks of gouty arthritis. Which is the exception? (UPSC)

1. Allopurinol
2. Aspirin

3. Probenecid
4. Febuxostat

DISCUSSION:
Aspirin, thiazide diuretics, niacin, and pyrazinamide are common drugs implicated in the origin of hyperuricemia or gout.

Drugs commonly used for preventing attacks of gout include xanthine oxidase inhibitors (such as allopurinol and febuxostat) and uricosuric drugs (such as probenecid).

ANSWER:
The correct answer is ‘aspirin’.

REFERENCES:
Schumacher H, Chen LX. Chapter 333. Gout and Other Crystal-Associated Arthropathies. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Zhang W, Doherty M, Bardin T, et al. (October 2006). EULAR evidence based recommendations for gout. Part II: Management. Report of a task force of the EULAR Standing Committee for International Clinical Studies Including Therapeutics (ESCISIT). Ann Rheum Dis 65 (10): 1312–24.

Question 58.
As per the current recommendations, the first line of treatment for ankylosing spondylitis is: (PGI)

1. NSAIDs
2. Golimumab
3. Etanercept
4. Infliximab

DISCUSSION:
Infliximab, adalimumab, certolizumab pegol, etanercept, and golimumab are together known as TNF

alpha inhibitors. Some of these agents have been found very useful in the treatment of ankylosing spondylitis. However, because they are expensive and have serious side effects, as per current recommendation, their use is restricted to patients with active disease that is inadequately responsive to therapy with at least two different NSAIDs.

Therefore, the first line of treatment for ankylosing spondylitis is NSAIDs.

ANSWER:
The correct answer is 'NSAIDs'.

REFERENCE:
Taurog JD. Chapter 325. The Spondyloarthritides. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 59.

Which one of the following features is not seen in reactive arthritis? (AIIMS)

1. Subcutaneous nodules
2. Keratoderma blennorrhagicum
3. Circinate balanitis
4. Sausage digit

DISCUSSION:
Reactive arthritis is an acute non-purulent arthritis complicating enteric or urogenital infections. The arthritis is usually asymmetric and mainly affects knee and ankle. Sausage digit or dactylitis is a manifestation of enthesopathy, a common feature of seronegative spondyloarthritides. Skin lesions include keratoderma blenorrhagica (mainly seen on palms and soles) and circinate balanitis (penis).

Subcutaneous nodules are found in rheumatoid arthritis.

ANSWER:
The correct answer is 'subcutaneous nodules'.

REFERENCE:

Kvien T, Glennas A, Melby K, Granfors K, et al. (1994). Reactive arthritis: Incidence, triggering agents and clinical presentation. J Rheum 21 (1): 115–22.

Question 60.

Which one of the following is a characteristic feature of Reiter's disease? (PGI)

1. Conjunctivitis
2. Subcutaneous nodules
3. Precipitated by streptococcal throat infection
4. Positive rheumatoid factor

DISCUSSION:

Reiter's disease is a type of reactive arthritis, and is characterized by the triad of arthritis, conjunctivitis, and urethritis. Reactive arthritis characteristically shows rheumatoid factor negativity, male predominance, onset usually before age 40, and association with HLA-B27. It is usually precipitated by enteric or urogenital infections.

Subcutaneous nodules are not seen in Reiter's disease.

ANSWER:

The correct answer is 'conjunctivitis'.

REFERENCE:

Braun J, Sieper J. Chapter 19.6. Ankylosing spondylitis, other spondyloarthritides, and related conditions. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 61.

All of the following conditions except one are frequently observed in gout. Which is the exception? (AIIMS)

1. Uric acid nephrolithiasis
2. Deficiency of enzyme xanthine oxidase
3. Hyperuricemia
4. Renal disease involving interstitial tissues

DISCUSSION:

Uric acid is the final breakdown product of purine metabolism. It is produced in tissues that contain enzyme xanthine oxidase. Enzyme defects associated with hyperuricemia include increased PRPP (phosphoribosylpyrophosphate) synthetase activity and deficiency of HPRT (hypoxanthine phosphoribosylpyrophosphate) activity.

Deficiency of xanthine oxidase does not produce hyperuricemia. In fact, xanthine oxidase inhibitors like allopurinol and febuxostat are used for controlling high serum uric acid levels.

ANSWER:

The correct answer is ‘deficiency of enzyme xanthine oxidase’.

REFERENCE:

Emmerson BT (1996). The management of gout. New Engl J Med, 334, 445–51.

Question 62.

The crystals isolated from joint in a patient with pseudogout consist of: (AIIMS)

1. Calcium pyrophosphate
2. Sodium urate
3. Potassium urate
4. Sodium pyrophosphate

DISCUSSION:

Calcium pyrophosphate dihydrate crystal deposition in cartilage of joints causes chondrocalcinosis. It is often asymptomatic, but can cause acute synovitis – also known as pseudogout (the term was coined by McCarty because of similarity with gout).

These crystals appear rhomboid, square, or rodlike under polarized light microscopy and show positive birefringence.

ANSWER:

The correct answer is ‘calcium pyrophosphate’.

REFERENCE:

Dieppe P, Swan A (1 May 1999). Identification of crystals in synovial fluid. Ann Rheum Dis 58 (5): 261–3.

RHEUMATOLOGY SECTION FOUR - AUTOIMMUNE DISORDERS

Question 63.

Which of the following is the most common clinically apparent cardiac manifestation of systemic lupus erythematosus? (AIIMS)

1. Myocarditis
2. Libman-Sacks endocarditis
3. Pericarditis
4. Aortic regurgitation

DISCUSSION:

The most common way in which lupus affects the heart is through inflammation of the pericardium, in the form of acute fibrinous pericarditis and pericardial effusion.

Though serious, lupus myocarditis and endocarditis are clinically less common. The endocarditis of SLE is characteristically noninfective (Libman-Sacks endocarditis), and can lead to thromboembolic phenomena. Aortic regurgitation is an infrequent manifestation of SLE.

ANSWER:

The correct answer is ‘pericarditis’.

REFERENCES:

Rahman A, Isenberg DA. Chapter 19.11.2. Systemic lupus erythematosus and related disorders. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Ansari A, Larson PH, Bates HD (May-June 1985). Cardiovascular manifestations of systemic lupus erythematosus: current perspective. Prog Cardiovasc Dis 27 (6): 421–34.

Question 64.

All of the following except one are typical clinical features of scleroderma. Which is the exception? (AI)

1. Dysphagia
2. Raynaud's phenomenon
3. Skin contracture
4. Malar rash

DISCUSSION:

Systemic sclerosis is a connective tissue disorder causing sclerodactyly and Raynaud's phenomenon, and affecting internal organs, notably the lungs (pulmonary artery hypertension, ILD), gastrointestinal tract (dysphagia), heart, and kidneys (hypertensive renal crisis).

Malar rash is a characteristic feature of systemic lupus erythematosus, and not systemic sclerosis.

ANSWER:

The correct answer is 'malar rash'.

REFERENCE:

Mayes MD, et al (2003). Prevalence, incidence, survival, and disease characteristics of systemic sclerosis in a large US population. Arthritis Rheum 48: 2246.

Question 65.

Which of the following is least likely to be seen as a manifestation of SLE? (PGI)

1. Pterygium
2. Alopecia
3. Anemia
4. Arthritis

DISCUSSION:
Alopecia, anemia, and arthritis are all common clinical features of SLE. Common ocular features of SLE include episcleritis, conjunctivitis, and the presence of cytoid bodies (white patches on the retina). However, pterygium is not a characteristic feature of SLE.

Pterygium (Surfer's Eye) is a benign growth of fibroblastic tissue on the nasal side of the sclera, often associated with chronic UV exposure.

ANSWER:
The correct answer is ‘pterygium’.

REFERENCES:
Hahn B. Chapter 319. Systemic lupus erythematosus. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Hahn B. Chapter 319. Systemic Lupus Erythematosus. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 66.

All of the following except one are typical clinical features of scleroderma. Which is the exception? (AI)

1. Pulmonary fibrosis
2. Esophageal dysmotility
3. Resorption of terminal phalanges
4. Lung nodules

DISCUSSION:
Resorption of the terminal phalanges (acro-osteolysis) can be seen in scleroderma. Other common causes include psoriasis, burn injury, frostbite, diabetic neuropathy, and hyperparathyroidism.

Esophageal dysmotility is seen in 80-90% of cases with systemic sclerosis. Pulmonary fibrosis is

more common in diffuse cutaneous systemic sclerosis than in limited cutaneous systemic sclerosis.

Lung involvement in scleroderma is in the form of lung fibrosis and pulmonary artery hypertension. Pulmonary nodules are found in conditions such as rheumatoid arthritis and granulomatosis with polyangiitis.

ANSWER:
The correct answer is ‘lung nodules’.

REFERENCE:
Mayes MD, et al (2003). Prevalence, incidence, survival, and disease characteristics of systemic sclerosis in a large US population. Arthritis Rheum 48: 2246.

Question 67.

All of the following except one are clinical features of systemic lupus erythematosus. Which is the exception? (PGI)

1. Recurrent abortion
2. Sterility
3. Coombs' positive hemolytic anemia
4. Psychosis

DISCUSSION:
Recurrent abortions are seen with anti-phospholipid syndrome. Coombs’ positive hemolytic anemia, signifying the presence of antibodies to red blood cells, is one of the diagnostic criteria for SLE. CNS lupus may show cognitive dysfunction, seizures, headache, psychosis, etc.

Patients with SLE may have recurrent abortions but not sterility (i.e., SLE does not reduce the ability to conceive).

ANSWER:
The correct answer is ‘sterility’.

REFERENCES:

Rahman A, Isenberg DA. Chapter 19.11.2. Systemic lupus erythematosus and related disorders. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Cervera R, et al (2002). Antiphospholipid syndrome: Clinical and immunologic manifestations and patterns of disease expression in a cohort of 1,000 patients. Arthritis Rheum 46: 1019.

Question 68.

Which one of the following antibodies is most useful in diagnosing Sjögren's syndrome? (AI)

1. Anti SS-A
2. Anti SCL-70
3. Anti-centromere
4. Anti-histone

DISCUSSION:

Anti SCL-70 antibody is often positive in diffuse and limited cutaneous scleroderma. Anti-centromere antibody is mainly seen in limited cutaneous scleroderma. Anti-histone antibody is mainly found in drug-induced lupus.

Anti SS-A (along with anti SS-B) is most useful in the diagnosis of Sjögren's syndrome. It is present in about two-thirds of affected patients.

ANSWER:

The correct answer is 'anti SS-A'.

REFERENCE:

Moutsopoulos HM, Tzioufas AG. Chapter 324. Sjögren's Syndrome. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 69.

All of the following except among the diagnostic criteria for SLE. Which is the exception? (UPSC)

1. Malar rash
2. Libman-Sacks endocarditis
3. Photosensitivity
4. Serositis

DISCUSSION:

Diagnostic criteria for SLE require the presence of any four of the following features: malar rash, discoid rash, photosensitivity, oral ulcers, serositis, renal disease, arthritis, ANA positivity, neurological disorders, hematological disorders, and presence of other (anti ds-DNA, anti-Sm, or antiphospholipid) antibodies.

Libman-Sacks endocarditis is not one of the diagnostic criteria.

ANSWER:

The correct answer is 'Libman-Sacks endocarditis'.

REFERENCES:

Hahn B. Chapter 319. Systemic lupus erythematosus. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Hochberg MC (1997). Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 40: 1725.

Question 70.

Which one of the following best describes the patient profile of Sjögren's syndrome? (AIIMS)

1. Male in 20-40 years age group
2. Female in 20-40 years age group
3. Male in 40-60 years age group

4. Female in 40-60 years age group

DISCUSSION:
Diagnosis of this condition requires the presence of four of the following six criteria: ocular signs or symptoms, oral signs or symptoms, and antibodies to SS-A/SS-B, and /or positive labial biopsy. In addition, presence of either positive biopsy or antibody is a must.

Sjögren's syndrome is mainly seen in middle-aged women (female-to-male ratio 9:1), although it may occur in all ages.

ANSWER:
The correct answer is 'female in 40-60 years age group'.

REFERENCE:
Venables PJW. Chapter 19.11.6. Sjögren's syndrome. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 71.

All of the following statements except one are correct about drug-induced SLE. Which is the exception? (AIIMS)

1. Anti-ds DNA antibodies are commonly found
2. Anti-histone antibodies are commonly found
3. CNS involvement not common
4. Renal involvement not common

DISCUSSION:
Drug-induced lupus appears during therapy with certain medications such as hydralazine, propylthiouracil, chlorpromazine, procainamide, and isoniazid. It usually resolves after discontinuation of the offending medication.

Drug-induced lupus differs from SLE in several aspects, namely in having almost equal sex

distribution, rarely involving kidneys or brain, being rarely associated with anti-ds DNA, and commonly associated with anti-histone antibodies.

ANSWER:
The correct answer is ‘anti-ds DNA antibodies are commonly found’.

REFERENCES:
Rahman A, Isenberg DA. Chapter 19.11.2. Systemic lupus erythematosus and related disorders. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Antonov D, Kazandjieva J, Etugov D, Gospodinov D, Tsankov N (Mar-April 2004). Drug-induced lupus erythematosus. Clin Dermatol 22 (2): 157–66.

Question 72.

Which of the following is an indicator of poor prognosis in a patient with systemic sclerosis? (AI)

1. Calcinosis cutis
2. Renal involvement
3. Dysphagia
4. Telangiectasia

DISCUSSION:
In terms of outcome, patients with the diffuse cutaneous form of scleroderma have a worse prognosis than those with the limited cutaneous form. Scleroderma renal crisis is an important cause of morbidity and mortality. About one-third of patients die within three years of developing renal crisis. Other important causes of death include pulmonary artery hypertension, pulmonary fibrosis, and gastrointestinal and cardiac complications.

ANSWER:
The correct answer is ‘renal involvement’.

REFERENCE:

Scussel-Lonzetti L, et al. (2002). Predicting mortality in systemic sclerosis: Analysis of a cohort of 309 French Canadian patients with emphasis on features at diagnosis as predictive factors for survival. Medicine (Baltimore) 81: 154.

Question 73.

Which one of the following clinical features is most likely to be seen in a patient with SLE? (PGI)

1. Uveitis
2. Joint deformity
3. Polyserositis
4. Cavitating lesion in lung

DISCUSSION:

Anterior uveitis is the most common extra-articular manifestation of ankylosing spondylitis. Cavitating lesion in the lung is characteristic of granulomatosis with polyangiitis (Wegner's). Joint deformity is not a feature of SLE, but seen in conditions such as rheumatoid arthritis and psoriatic arthritis.

Polyserositis is seen in SLE and usually manifests are pleuritis or pericarditis.

ANSWER:

The correct answer is 'polyserositis'.

REFERENCES:

Hahn B. Chapter 319. Systemic lupus erythematosus. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Rahman A, Isenberg D (2008). Systemic lupus erythematosus. New Engl J Med 358: 929.

Question 74.

Which one of the following conditions is most likely to be associated with Sjögren's syndrome? (PGI)

1. Rheumatoid arthritis
2. Scleroderma
3. SLE
4. Mixed connective tissue disorder

DISCUSSION:

There are two types of Sjögren's syndrome. Primary Sjögren's syndrome is not associated with other autoimmune or connective tissue disorders. Secondary Sjögren's syndrome is associated with other disorders such as rheumatoid arthritis, scleroderma, mixed connective tissue disorder, and polymyositis.

Of these, rheumatoid arthritis is the most common cause of secondary Sjögren's syndrome.

ANSWER:

The correct answer is 'rheumatoid arthritis'.

REFERENCE:

Garcia-Carrasco M, et al. (2006). Pathophysiology of Sjögren's syndrome. Arch Med Res 37: 921.

Question 75.

All of the following factors except one are associated with adverse prognosis and high risk of renal progression in lupus nephritis. Which is the exception? (PGI)

1. High levels of anti-ds DNA
2. Persistent proteinuria (nephrotic range > 3gm/day)
3. Hypocomplementenemia
4. Anti La (SS-B)

DISCUSSION:
Diagnosis of lupus nephritis is suggested with urine analysis, as patients are often asymptomatic, but classification requires biopsy. A renal lupus flare is indicated by an increase in proteinuria and/or serum creatinine concentration, abnormal urine sediment, or a reduction in creatinine clearance rate as a result of active disease. Other predictors of flare rising levels of anti-ds DNA are reducing levels of complements C3 or C4.

Anti Ro (SS-A) and anti La (SS-B) antibodies are associated with reduced risk of lupus nephritis.

ANSWER:
The correct answer is 'anti La (SS-B)'.

REFERENCES:
Hahn B. Chapter 319. Systemic lupus erythematosus. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Sprangers BI, Monahan M, Appel GB (Dec 2012). Diagnosis and treatment of lupus nephritis flares--an update. Nat Rev Nephrol 8 (12): 709–17.

Question 76.

The most specific antibody in the diagnosis of SLE is: (PGI)

1. ANA
2. Anti-RNP
3. Anti-ss DNA
4. Anti-Sm

DISCUSSION:
The best screening test for SLE is the antinuclear antibody (ANA) assay. ANA is highly sensitive with over 95% of patients with SLE, but the specificity is not high. Anti-RNP antibodies are detectable in some SLE patients, but high titers of anti-RNP antibodies are diagnostic of MCTD. Anti-ss DNA (single stranded DNA) is non-specific and rarely indicated.

Anti-ds DNA (double stranded DNA) and anti-Sm antibodies are considered the most specific tests for SLE.

ANSWER:
The correct answer is ‘anti-Sm’.

REFERENCES:
Rahman A, Isenberg DA. Chapter 19.11.2. Systemic lupus erythematosus and related disorders. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Hahn B. Chapter 319. Systemic Lupus Erythematosus. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 77.

Which one of the following statements is true in considering treatment of systemic sclerosis? (AI)

1. Nifedipine is used for Raynaud’s phenomenon
2. ACE inhibitors are contraindicated in scleroderma
3. Systemic steroids are useful for treating skin changes
4. There are no effective oral treatments for pulmonary hypertension

DISCUSSION:
Systemic steroids are avoided as far as possible, as they have been implicated in precipitating hypertensive renal crisis in scleroderma patients. Moreover, they have not been shown to improve skin changes in scleroderma. Bosentan is an oral endothelin antagonist, used for pulmonary hypertension. Short acting ACE inhibitors are indicated for rapid blood pressure control during scleroderma renal crisis.

ANSWER:
The correct answer is ‘nifedipine is used for Raynaud’s phenomenon’.

REFERENCE:

Varga J. Chapter 323. Systemic Sclerosis (Scleroderma) and Related Disorders. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 78.

Antinuclear antibodies are present in what percentage of cases of SLE? (UPSC)

1. Less than 30%
2. 50%
3. 50–70 %
4. More than 90%

DISCUSSION:

The antinuclear antibodies are considered the best screening test for SLE. Their absence makes the diagnosis of SLE unlikely. ANA is highly sensitive with over 95% of patients with SLE who are ANA positive, but the specificity is not high. Anti-ds DNA (double stranded DNA) and anti-Sm antibodies are considered the most specific tests for SLE.

ANSWER:

The correct answer is ‘more than 90%’.

REFERENCES:

Hahn B. Chapter 319. Systemic lupus erythematosus. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Kumar Y, Bhatia A, Minz RW (2 Jan 2009). Antinuclear antibodies and their detection methods in diagnosis of connective tissue diseases: a journey revisited. Diagnostic Pathology 4: 1.

Question 79.

Which of the following statements are correct about Sjögren's syndrome? (AIIMS)

1. Glomerulonephritis is common
2. RA factor is negative
3. May be associated with lymphoma
4. Antinuclear antibodies are negative

DISCUSSION:

Sjögren's syndrome is an autoimmune disorder affecting mainly the salivary and lacrimal glands. Lymphocytic infiltration of these glands leads to dry eyes and dry mouth. Sjögren's syndrome is associated with rheumatoid factor in up to 75% of patients, although rheumatoid arthritis is less frequently present. Similarly, antinuclear antibodies are found in more than 80% of patients. Tubulo-interstitial disorders are more common in the kidneys, while glomerulonephritis is uncommon.

Lymphoma may be seen in up to 5% of patients with Sjögren's syndrome.

ANSWER:

The correct answer is 'may be associated with lymphoma'.

REFERENCE:

Fox RI (2005). Sjögren's syndrome. Lancet 366, 321–331.

Question 80.

Which of the following antibodies is found most commonly in a patient with SLE? (AIIMS)

1. Antinuclear antibody
2. Anti-ds DNA antibody
3. Anti-Sm antibody
4. Anti-histone antibody

DISCUSSION:
Antinuclear antibody is found in >95% of cases of SLE, and is considered the best screening test. Anti-ds DNA and anti-Sm antibodies are considered specific, but are found less frequently. Anti-histone antibodies are found in about 95% of cases of drug-induced lupus, but in 70% of cases of SLE.

ANSWER:
The correct answer is ‘antinuclear antibody’.

REFERENCES:
Rahman A, Isenberg DA. Chapter 19.11.2. Systemic lupus erythematosus and related disorders. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Hahn B. Chapter 319. Systemic lupus erythematosus. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 81.
All of the following except one are typical features of diffuse cutaneous scleroderma. Which is the exception? (PGI)

1. Raynaud's phenomenon
2. Trunk involvement
3. Anti-centromere antibodies are characteristic
4. Associated with hypertensive renal crisis

DISCUSSION:
Diffuse cutaneous scleroderma has higher chances of developing pulmonary fibrosis and hypertensive renal crisis than limited cutaneous scleroderma. Raynaud’s phenomenon is present in both diffuse cutaneous and limited cutaneous scleroderma. Trunk involvement is seen only in diffuse cutaneous scleroderma.

Anti-centromere antibodies are characteristic of limited cutaneous (and not diffuse cutaneous)

scleroderma.

ANSWER:

The correct answer is ‘anti-centromere antibodies are characteristic’.

REFERENCE:

Varga J, Abraham D (2007). Systemic sclerosis: A prototypic multisystem fibrotic disorder. J Clin Invest 117: 557.

RHEUMATOLOGY SECTION FIVE - MISCELLANEOUS RHEUMATOLOGY

Question 82.

Anti-topoisomerase I antibody is a marker of which of the following diseases? (JIPMER)

1. Systemic sclerosis
2. Classic polyarteritis nodosa
3. Systemic lupus erythematosus
4. Rheumatoid arthritis

DISCUSSION:

Anti-topoisomerase I antibodies, also known as anti-Scl 70 antibodies are seen mainly in 'diffuse cutaneous' systemic sclerosis. However, they are also seen (less commonly) in 'limited cutaneous' systemic sclerosis. Another antibody that can be seen in the 'diffuse cutaneous' form is anti-RNA polymerase III antibody. Anti-centromere antibodies are found in the 'limited cutaneous' form of scleroderma.

Anti-CCP antibodies and RA factor are useful in rheumatoid arthritis. ANA (antinuclear antibodies) are a good screening test for SLE. ANCA is usually negative in classical polyarteritis nodosa.

ANSWER:

The correct answer is 'systemic sclerosis'.

REFERENCE:

Hinchcliff M, Varga J (October 2008). Systemic sclerosis/scleroderma: a treatable multisystem disease. Am Fam Physician 78 (8): 961–8.

Question 83.

Upper respiratory tract infections are common in which one of the following diseases? (PGI)

1. Granulomatosis with polyangiitis
2. Polyarteritis nodosa
3. Systemic lupus erythematosus
4. Giant cell arteritis

DISCUSSION:

Granulomatosis with polyangiitis is an important cause of ANCA-associated vasculitis. It is characterized by the presence of granulomatous vasculitis of upper and lower respiratory tract, along with glomerulonephritis. Sinusitis, otitis media, and nasal purulent discharge are all common presentations of upper respiratory tract involvement in these patients.

Polyarteritis nodosa and giant cell arteritis do not involve usually affect the respiratory tract. The most common pulmonary involvement in systemic lupus erythematosus is in the form of pleuritis and pleural effusion.

ANSWER:

The correct answer is ‘granulomatosis with polyangiitis’.

REFERENCE:

Berden A, Göçeroglu A ,et al. (Jan 2012). Diagnosis and management of ANCA associated vasculitis. BMJ 16: 344:e26.

Question 84.

All of the following except one frequently show bony erosions. Which is the exception? (AIIMS)

1. Gout
2. Psoriasis
3. SLE
4. Rheumatoid arthritis

DISCUSSION:

The term 'bony erosions' refers to loss of mineralized bone at juxta-articular sites. Determination of bone erosion is almost exclusively based on radiographic findings, and is characteristic of rheumatoid arthritis and other chronic forms of arthritis, such as gouty and psoriatic arthritis.

Arthralgia/arthritis is the most common symptom in SLE. However, joint deformities and erosions are rare. When joint deformities do occur, they are secondary to tenosynovitis, rather than to joint damage.

ANSWER:

The correct answer is ‘SLE’.

REFERENCES:

Hahn B. Chapter 319. Systemic lupus erythematosus. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Zoma A (2004). Musculoskeletal involvement in systemic lupus erythematosus. Lupus 13: 851–3.

Question 85.

A 38-year-old male suffered from redness and swelling of the first metatarsophalangeal joint three times over the past two years. He was found to be hypertensive, and the EEG showed inverted 'T' waves in precordial leads. Which one of the following investigations will be diagnostic? (UPSC)

1. Rheumatoid factor
2. Serum uric acid
3. ESR
4. Joint fluid aspiration

DISCUSSION:

The description provided is that of recurrent attacks of gout. Definitive diagnosis of an acute attack of gout requires identification of monosodium urate crystals in the joint fluid aspirate. During an acute attack, the ESR is frequently elevated, but it is a non-specific finding. The uric acid level may or may

not increase during acute attack, and does not help in making diagnosis of gout. Typically, patients with rheumatoid arthritis do not develop gout.

A hypertensive patient with ECG showing 'T' wave inversion is likely to be receiving aspirin, which can at times produce hyperuricemia.

ANSWER:
The correct answer is 'joint fluid aspiration'.

REFERENCES:
Edward Roddy E, Doherty M. Chapter 19.10. Crystal-related arthropathies. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Underwood M (2006). Diagnosis and management of gout. Br Med J, 332, 1315–19.

Question 86.

Shrinking lung is a feature of: (PGI)

1. SLE
2. Rheumatoid arthritis
3. Scleroderma
4. Sarcoidosis

DISCUSSION:
Shrinking lung syndrome is a rare complication of systemic lupus erythematosus. It is seen during the later stage of the disease and presents with dyspnea and pleuritic chest pain. Investigations reveal elevated hemidiaphragm and restriction on pulmonary function tests. It is treated with steroids and prognosis is good.

ANSWER:
The correct answer is 'SLE'.

REFERENCE:
Singh RI, Huang W, Menon Y, Espinoza LR (Dec 2002). Shrinking lung syndrome in systemic lupus erythematosus and Sjögren's syndrome. J Clin Rheumatol 8 (6): 340–5.

Question 87.

Which of the following is correct about Behçet's disease? (AIIMS)

1. There is a strong association with HLA-B7
2. The skin may be hyperactive to minor injury such as venepuncture
3. Renal manifestation
4. Lung involvement

DISCUSSION:
In patients with Behçet's disease, a minor trauma such as venepuncture often leads to the development of skin lesions or ulcers that may be resistant to healing. This is called a pathergy reaction, and is considered a unique feature of Behçet's disease.

Behçet's disease may be associated with HLA-B51. Kidney and lung involvement is not seen in Behçet's disease.

ANSWER:
The correct answer is 'the skin may be hyperactive to minor injury such as venepuncture'.

REFERENCE:
Geri G, et al. (Jan 2012). Spectrum of cardiac lesions in Behçet disease: a series of 52 patients and review of the literature. Medicine (Baltimore) 91 (1): 25–34.

Question 88.

Systemic lupus erythematosus occurs most commonly in: (AIIMS)

1. Children
2. Elderly men
3. Elderly women
4. Women of childbearing age

DISCUSSION:
Overall, more than 80% of patients with systemic lupus erythematosus are women. The majority of them are of childbearing age. This has been attributed to the possible role played by sex hormones in the pathogenesis of this disease (most cases develop after menarche and before menopause).

ANSWER:
The correct answer is 'women of childbearing age'.

REFERENCES:
Rahman A, Isenberg DA. Chapter 19.11.2. Systemic lupus erythematosus and related disorders. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Cooper GS, Dooley MA, Treadwell EL, St Clair EW, Parks CG, Gilkeson GS (Oct 1998). Hormonal, environmental, and infectious risk factors for developing systemic lupus erythematosus. Arthritis Rheum 41 (10): 1714–24.

Question 89.
Anti-ds DNA antibody is highly specific for: (JIPMER)

1. Systemic sclerosis
2. SLE
3. Polymyositis
4. Multiple sclerosis

DISCUSSION:
Anti-ds DNA (and anti-Sm) antibodies are considered the most specific tests for SLE. In some

patients, anti- double stranded DNA correlates with disease severity.

The best screening test for SLE is the ANA (antinuclear antibody) assay.

ANSWER:
The correct answer is ‘SLE’.

REFERENCES:
Rahman A, Isenberg DA. Chapter 19.11.2. Systemic lupus erythematosus and related disorders. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Rahman A, Isenberg DA. Chapter 19.11.2. Systemic lupus erythematosus and related disorders. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 90.

Anti-histone antibodies are characteristics of which of the following conditions? (PGI)

1. Drug-induced lupus
2. Cardiac lupus
3. Lupus nephritis
4. MCTD

DISCUSSION:
No specific antibodies identify cardiac lupus. Predictors of lupus nephritis flare include rising levels of anti-ds DNA and reducing levels of complements C3 or C4. Anti-Ro (SS-A) and anti-La (SS-B) antibodies are associated with a reduced risk of lupus nephritis. Anti-RNP antibodies are detectable in less than half of SLE patients, but high titers of anti-RNP antibodies are diagnostic of MCTD.

Anti-histone antibodies are autoantibodies that are found in two-thirds of patients with systemic lupus erythematosus (SLE) and in more than 95% of patients with drug-induced lupus.

ANSWER:

The correct answer is ‘drug-induced lupus’.

REFERENCE:

Epstein A, Barland P (Feb 1985). The diagnostic value of antihistone antibodies in drug-induced lupus erythematosus. Arthritis Rheum 28 (2): 158–62.

Question 91.

All of the following except one are features of Felty’s syndrome. Which is the exception? (PGI)

1. Rheumatoid arthritis
2. Splenomegaly
3. Neutropenia
4. Osteoarthritis

DISCUSSION:

Felty’s syndrome is named for Augustus Roi Felty. It is described as a clinical triad of neutropenia, splenomegaly, and rheumatoid arthritis. Lymphadenopathy, leg ulcers, nodular hyperplasia of the liver, and anemia are some other features seen in these patients.

Osteoarthritis is not a feature of Felty’s syndrome.

ANSWER:

The correct answer is ‘Felty’s syndrome’.

REFERENCES:

Shah A, St. Clair E. Chapter 321. Rheumatoid Arthritis. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Balint GP, Balint PV (Oct 2004). Felty's syndrome. Best Pract Res Clin Rheumatol 18 (5): 631–645.

Question 92.

In all except one of the following diseases, enthesitis is a prominent feature. Which is the exception? (JIPMER)

1. Reiter's disease
2. Psoriatic arthritis
3. Rheumatoid arthritis
4. Ankylosing spondylitis

DISCUSSION:

The spondyloarthritides mainly include ankylosing spondylitis, reactive arthritis, psoriatic arthritis, and enteropathic arthritis. These disorders share some common features, including a striking association with HLA-B27, male predominance, onset usually before age 40, inflammatory arthritis of the spine and sacroiliac joints, and the absence of autoantibodies in the serum.

Enthesitis (inflammation at tendon, ligament, or joint capsule insertions) is a characteristic feature of spondyloarthritides, and not rheumatoid arthritis.

ANSWER:

The correct answer is ‘rheumatoid arthritis’.

REFERENCE:

Braun J, Sieper J. Chapter 19.6. Ankylosing spondylitis, other spondyloarthritides, and related conditions. In: Warrell DA, Cox TM, Firth JD, eds. Oxford Textbook of Medicine. 5th edition.

Question 93.

Which of the following is an example of small vessel vasculitis? (PGI)

1. Takayasu’s arteritis
2. Polyarteritis nodosa
3. Kawasaki disease
4. Granulomatosis with polyangiitis

DISCUSSION:
Large vessel vasculitides are giant cell arteritis, Takayasu's arteritis, and isolated CNS angiitis. Medium vessel vasculitides are PAN and Kawasaki disease (in childhood).

Granulomatosis with polyangiitis is an example of small vessel vasculitis. Other examples of small vessel vasculitis are microscopic polyangiitis, Churg–Strauss syndrome, Henoch–Schönlein purpura, and cryoglobulinemic vasculitis.

ANSWER:
The correct answer is ‘granulomatosis with polyangiitis’.

REFERENCE:
Jennette J, et al. (2013). 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Arthritis Rheum 65, 1–11.

Question 94.

Martel's sign is seen in which of the following conditions? (APPGE)

1. Rheumatoid arthritis
2. Ankylosing spondylitis
3. Gout
4. Osteoarthritis

DISCUSSION:
Martel’s sign, also known as rat bite sign, is an X-ray finding seen in patients with gout. It refers to well-defined, ‘punched-out’ type bony erosions with sclerotic margins, resulting from a long-standing, soft-tissue tophus in patients with gout.

ANSWER:
The correct answer is ‘gout’.

REFERENCE:
www.japi.org/september2005/PC-782.pdf.

http://medical-dictionary.thefreedictionary.com/rat bite sign

Question 95.

All of the following except one are examples of small vessel vasculitis. Which is the exception? (AI)

1. Granulomatosis with polyangiitis
2. Microscopic polyangiitis
3. Polyarteritis nodosa
4. Henoch-Schönlein purpura

DISCUSSION:
Granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis (Churg–Strauss), microscopic polyangiitis, anti-glomerular basement membrane (GBM) disease, IgA vasculitis (Henoch–Schönlein), and cryoglobulinemic-associated vasculitides predominantly affect small-sized vessels.

Polyarteritis nodosa and Kawasaki disease-associated vasculitides predominantly affect medium-sized vessels.

ANSWER:
The correct answer is ‘polyarteritis nodosa’.

REFERENCE:
Jennette J, et al. (2013). 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Arthritis Rheum 65, 1–11.

Question 96.

All of the following medical conditions except one are associated with osteoporosis. Which is the exception? (AIIMS)

1. Thyrotoxicosis
2. Rheumatoid arthritis
3. Hypoparathyroidism
4. Steroid therapy

DISCUSSION:

The most common causes of osteoporosis include post-menopausal status, aging, high-dose corticosteroid administration, alcoholism, smoking, and sex hormone deficiency. Chronic inflammatory disorders such as rheumatoid arthritis are also associated with osteoporosis.

Common endocrine causes associated with osteoporosis include Cushing's syndrome, thyrotoxicosis, and hyperparathyroidism (not hypoparathyroidism). In fact, an increased bone density has been reported in patients with hypoparathyroidism.

ANSWER:

The correct answer is ‘ hypoparathyroidism’.

REFERENCE:

Touliatos JSI, Sebes JI, Hinton A, McCommon D, Karas JG, Palmieri GM (Aug 1995). Hypoparathyroidism counteracts risk factors for osteoporosis. Am J Med Sci 310 (2): 56–60.

Question 97.

All of the following except one are also known as ‘ANCA’-associated vasculitides. Which is the exception? (PGI)

1. Granulomatosis with polyangiitis
2. Churg–Strauss disease
3. Microscopic polyangiitis

4. Takayasu's arteritis

DISCUSSION:
Granulomatosis with polyangiitis (Wegner's), Churg–Strauss disease, and microscopic polyangiitis are also known as ANCA-associated vasculitis. Cytoplasmic ANCA (cANCA) mainly detects proteinase-3, while perinuclear ANCA (pANCA) mainly detects myeloperoxidase enzyme.

Takayasu's arteritis is also known as 'aortic arch syndrome' and the 'pulseless disease'. Here, antineutrophil cytoplasmic antibodies are as common as in the general population.

ANSWER:
The correct answer is 'Takayasu's arteritis'.

REFERENCE:
Savige J, Davies D, Falk RJ, Jennette JC, Wiik A (Mar 2000). Antineutrophil cytoplasmic antibodies and associated diseases: a review of the clinical and laboratory features. Kidney International 57 (3): 846–62.

Question 98.

In a patient with suspected vasculitis, the 'cANCA' is most likely to be seen in which one of the following conditions? (AIIMS)

1. Polyarteritis nodosa
2. Granulomatosis with polyangiitis
3. Henoch–Schönlein purpura
4. Churg–Strauss syndrome

DISCUSSION:
Anti-neutrophil cytoplasmic antibodies (ANCAs) are mainly IgG antibodies against antigens in the cytoplasm of granulocytes. Two common patterns are described by immunofluorescence: 'cANCA' (cytoplasmic ANCA) and 'pANCA' (perinuclear ANCA). Cytoplasmic ANCA (cANCA) mainly

detects proteinase-3, while perinuclear ANCA (pANCA) mainly detects myeloperoxidase enzyme.

More than 90% of patients with granulomatosis with polyangiitis have ‘cANCA’.

ANSWER:
The correct answer is ‘granulomatosis with polyangiitis’.

REFERENCE:
Bosch X, Guilabert A, Font J (2006). Antineutrophil cytoplasmic antibodies. Lancet, 368, 404–18.

Question 99.

All of the following except one are associated with Behçet’s disease. Which is the exception? (AIIMS)

1. Myocarditis
2. Erythema nodosum
3. Oral and genital ulcers
4. Thrombophlebitis

DISCUSSION:
Behçet's disease is a multisystem disorder presenting with recurrent oral and genital ulcerations, acneiform lesions, erythema nodosum, pathergy reaction, uveitis, arthritis, and thrombophlebitis.

Myocarditis is not a feature of Behçet’s disease.

ANSWER:
The correct answer is ‘myocarditis’.

REFERENCE:
Geri G, et al. (Jan 2012). Spectrum of cardiac lesions in Behçet disease: a series of 52 patients and review of the literature. Medicine (Baltimore) 91 (1): 25–34.

Question 100.

All of the following except one are features of seronegative spondyloarthropathy. Which is the exception? (PGI)

1. Strong association with HLA-B27
2. Negative rheumatoid factor
3. Symmetrical polyarthritis
4. Enthesitis

DISCUSSION:

The spondyloarthritides mainly include ankylosing spondylitis, reactive arthritis, psoriatic arthritis, and enteropathic arthritis. These disorders share some common features, including a striking association with HLA-B27, male predominance, onset usually before age 40, inflammatory arthritis of the spine and sacroiliac joints, and the absence of autoantibodies in the serum.

Symmetrical polyarthritis is not a feature of spondyloarthritides. They are characterized by asymmetric oligoarthritis of large peripheral joints.

ANSWER:

The correct answer is ‘symmetrical polyarthritis’.

REFERENCE:

Braun J, Sieper J (2007). Ankylosing spondylitis. Lancet, 369, 1379–90.

Question 101.

In a patient with suspected vasculitis, c-ANCA positivity indicates an antibody formed against: (AI)

1. Proteinase 3
2. Myeloperoxidase
3. Lactoferrin
4. Elastase

DISCUSSION:
Granulomatosis with polyangiitis (Wegner's), Churg–Strauss disease, and microscopic polyangiitis are also known as ANCA-associated vasculitis. Two common patterns are described by immunofluorescence: 'cANCA' (cytoplasmic ANCA) and 'pANCA' (perinuclear ANCA). The perinuclear ANCA (pANCA) mainly detects myeloperoxidase enzyme. Less frequent 'pANCA' targets include lactoferrin, elastase, and cathepsin G.

Cytoplasmic ANCA (cANCA) mainly detects proteinase-3.

ANSWER:
The correct answer is 'proteinase-3'.

REFERENCE:
Berden A, Göçeroglu A, et al. Jan 2012). Diagnosis and management of ANCA associated vasculitis. BMJ 16: 344:e26.

Question 102.

Which one of the following is a marker for bone formation? (AIIMS)

1. Tartrate resistant acid phosphatase
2. Osteocalcin
3. Urinary calcium
4. Urinary hydroxyproline

DISCUSSION:
Biochemical markers of bone formation are serum bone-specific alkaline phosphatase, serum osteocalcin, and serum type I procollagen.

The biochemical markers of bone resorption are urine and serum cross-linked N-telopeptide, urine and serum cross-linked C-telopeptide, tartarate resistant acid phosphatase, urine total free deoxypyridinoline, and urinary hydroxyproline.

ANSWER:
The correct answer is ‘osteocalcin’.

REFERENCE:
Lewiecki EM, Baim S, Bilezikian JP, Eastell R, LeBoff MS, Miller PD (Apr-Jun 2009). 2008 Santa Fe Bone Symposium: update on osteoporosis. J Clin Densitom 12 (2): 135–57.

Question 103.

A combination of cavitary lesions in lung, chronic sinusitis, and glomerulonephritis best describes which one of the following diseases? (PGI)

1. Granulomatosis with polyangiitis
2. Polyarteritis nodosa
3. Systemic lupus erythematosus
4. Goodpasture syndrome

DISCUSSION:
Polyarteritis nodosa does not involve pulmonary arteries. Most common pulmonary involvement in systemic lupus erythematosus is in the form of pleuritis and pleural effusion. Sinusitis is not a feature of Goodpasture syndrome. This patient is likely to have Wegner’s granulomatosis.

It is characterized by presence of granulomatous vasculitis of upper and lower respiratory tract. Kidney involvement may take the form of rapidly progressive glomerulonephritis.

ANSWER:
The correct answer is ‘granulomatosis with polyangiitis’.

REFERENCE:
Berden A, Göçeroglu A, et al. (Jan 2012). Diagnosis and management of ANCA associated vasculitis. BMJ 16; 344:e26.

Question 104.

A 50-year-old patient wakes up with excruciating pain at the base of the great toe, with redness and swelling. Which of the following would be the most desirable drug in this patient? (UPSC)

1. Allopurinol
2. Probenecid
3. Febuxostat
4. Indomethacin

DISCUSSION:

To manage an acute attack of gout, NSAIDs or colchicine are typically used. Other options include joint fluid aspiration and local injection of steroids.

Allopurinol and febuxostat are xanthine oxidase inhibitors, which are not started during an acute attack (sudden lowering of uric acid is not desirable). Similarly, the euricosuric drug probenecid should to be avoided during an acute attack.

ANSWER:

The correct answer is 'indomethacin'.

REFERENCES:

Schumacher H, Chen LX. Chapter 333. Gout and Other Crystal-Associated Arthropathies. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Underwood M (2006). Diagnosis and management of gout. Br Med J 332, 1315–19.

Question 105.

A 40-year-old woman presented with dysphagia and stiff fingers for the last three to four years. Which of the following conditions is the most likely cause? (AIIMS)

1. Systemic lupus erythematosus

2. Rheumatoid arthritis
3. Systemic sclerosis
4. Osteoarthritis

DISCUSSION:
Raynaud's phenomenon, dysphagia, and sclerodactyly are characteristic features of systemic sclerosis. Gastro-esophageal reflux is common in these patients and responds well to proton pump inhibitors. Raynaud's phenomenon may be helped by nifedipine.

Dysphagia is not a feature of rheumatoid arthritis, SLE, or osteoarthritis.

ANSWER:
The correct answer is 'systemic sclerosis'.

REFERENCE:
Rose S, Young MA, Reynolds JC (1998). Gastrointestinal manifestations of scleroderma. Gastroenterol Clin North Am. 27 (3): 563–94.

Question 106.

Which one of the following is associated with CREST (calcinosis, Raynaud's phenomenon, esophageal hypomotility, sclerodactyly, and telangiectasia) syndrome? (UPSC)

1. Systemic lupus erythematosus
2. Dermatomyositis
3. Polyarteritis nodosa
4. Systemic sclerosis

DISCUSSION:
The CREST syndrome is seen in some patients with limited cutaneous systemic sclerosis. It is characterized by the presence of anti-centromere antibodies. Patients with limited cutaneous systemic sclerosis have relatively limited involvement of skin, often confined to fingers, forearms, and face.

Notably, the trunk is spared. Their overall prognosis is better than those with diffuse cutaneous systemic sclerosis, mainly because of less visceral involvement.

ANSWER:
The correct answer is ‘systemic sclerosis’.

REFERENCE:
Winterbauer RH (1964). Multiple telangiectasia, Raynaud's phenomenon, sclerodactyly, and subcutaneous calcinosis: a syndrome mimicking hereditary hemorrhagic telangiectasia. Bulletin of the Johns Hopkins Hospital 114: 361–83.

Question 107.

HLA-B27 is a genetic marker for which of the following disorders? (AI)

1. Systemic lupus erythematosus
2. Behçet’s disease
3. Rheumatoid arthritis
4. Seronegative spondyloarthritis syndrome

DISCUSSION:
SLE is associated with mainly HLA-DR3, while rheumatoid arthritis is associated with HLA-DR4. Behçet’s disease is associated with HLA-B51.

Seronegative spondyloarthritis syndrome refers to a group of inflammatory rheumatic diseases with common features, including axial and peripheral inflammatory arthritis, enthesopathy, extra-articular manifestations, and a close association with HLA-B27. Diseases include ankylosing spondylitis, reactive arthritis, psoriatic arthritis, and enteropathic arthritis.

ANSWER:
The correct answer is ‘seronegative spondyloarthritis syndrome’.

REFERENCE:
Caillat-Zucman S (2008). Molecular mechanisms of HLA association with autoimmune diseases. Tissue Antigens 73: 1.

Question 108.

Schober's test is performed to know about: (PGI)

1. Flexion of lumbar spine
2. Chest expansion
3. Pain with motion of hip joint
4. Neck pain and stiffness

DISCUSSION:
Schober's test is performed to assess the mobility of the spine in ankylosing spondylitis. In this test, the patient stands erect with normal posture. Two points are marked at predefined landmarks over the back. The patient is then asked to bend forward. Ideally, the distance between two points should increase beyond a certain extent. Failure to do so suggests reduced flexion of lumbar spine and favors the diagnosis of ankylosing spondylitis.

ANSWER:
The correct answer is ‘flexion of lumbar spine’.

REFERENCE:
Braun J, Sieper J (2007). Ankylosing spondylitis. Lancet 369, 1379–90.

Question 109.

Keratoderma blennorrhagica is pathognomic of: (PGI)

1. Behçet's disease

2. Reiter's disease
3. Lyme's disease
4. Glucagonoma

DISCUSSION:
Reiter's disease is a type of reactive arthritis. Reactive arthritis shares certain clinical features with other seronegative spondyloarthritides. These include male predominance, onset usually before age 40, inflammatory arthritis of the spine and sacroiliac joints, asymmetric oligoarthritis, and a striking association with HLA-B27.

Keratoderma blennorrhagica are painless skin lesions (vesico-pustular waxy lesions with a yellow-brown color) commonly found on the palms and soles of patients with Reiter's disease.

ANSWER:
The correct answer is 'Reiter's disease'.

REFERENCE:
Tonna I, Laing RB (2008). Keratoderma Blennorrhagica. N Engl J Med 358: 2160.

Question 110.

All of the following conditions except one are more common in females. Which is the exception? (PGI)

1. Ankylosing spondylitis
2. Heberden's nodes
3. Rheumatoid arthritis
4. Systemic lupus erythematosus

DISCUSSION:
Heberden's nodes (named after William Heberden) are swellings that develop in the distal interphalangeal joints in patients with osteoarthritis. They are more common in women than in men.

Rheumatoid arthritis and systemic lupus erythematosus are typically more common in females.

Ankylosing spondylitis is a type of spondyloarthritis. Other types of spondyloarthritides include reactive arthritis, psoriatic arthritis, and enteropathic arthritis. These disorders show male predominance.

ANSWER:
The correct answer is ‘ankylosing spondylitis’.

REFERENCE:
Taurog JD. Chapter 325. The spondyloarthritides. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson J, Loscalzo J. eds. Harrison's Principles of Internal Medicine, 18th edition.

Question 111.

All of the following except one are features of Felty’s syndrome. Which is the exception? (PGI)

1. Splenomegaly
2. Rheumatoid arthritis
3. Leukocytosis
4. Lymphadenopathy

DISCUSSION:
The triad of splenomegaly, rheumatoid arthritis, and neutropenia (and not leukocytosis) describes Felty’s syndrome, named for Augustus Roi Felty.

Lymphadenopathy, leg ulcers, nodular hyperplasia of the liver, and anemia are some other features seen in these patients.

ANSWER:
The correct answer is ‘leukocytosis’.

REFERENCE:

Balint GP, Balint PV (Oct 2004). Felty's syndrome. Best Pract Res Clin Rheumatol 18 (5): 631–645.

Table of Contents